

Commissioning Tertiary and Specialised Services for Children and Young People

May 2004



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Royal College of Paediatrics and Child Health

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Foreword

Children have never been healthier, but for those who do need medical care there is a great deal more that can be done. In addition parental expectations of what should be done have never been higher. Our principle must be that every child deserves the best possible care and that this should be delivered as close to the child's own home as possible. Many specialised services have been developed in tertiary centres. There has been a move in recent years to a shared care arrangement where some aspects of care are delivered in the child's locality and only those which can not be delivered there are given in a more specialist centre. Managed networks involving shared care are of enormous value to the child. They ensure parents that they will have access to the best care in the right place by the best team.

The principle of devolution of management embodied in *Shifting the Balance of Power* means that Primary Care Organisations in England and similar structures in the other devolved countries of the UK now have to commission services. However, many problems in childhood are relatively uncommon so that comprehensive services can not be economically planned or commissioned at a local level.

The RCPCH have, therefore, produced this report which details the specialist and tertiary services that might be required. We hope that it will be of value to the National Service Framework for Children in England and to others who are planning children's services. However, it is not an end in itself and further work is continuing to define the best service for children.

Finally I would like to thank John Jenkins for masterminding the production of this report in a remarkably short period of time.

Professor Alan Craft
President, RCPCH
May 2004

Introduction

Throughout the United Kingdom health services are now mainly commissioned by primary care organisations (PCOs) on behalf of their local populations. It has been recognized that single PCOs, covering relatively small populations, will be unable to effectively commission services for rare conditions or highly complex treatments or care packages. Although children comprise over 20% of the population, many specific conditions affecting children are of relatively low prevalence or require high levels of specialist skill during treatment due to the physiological needs of smaller children and infants.

Guidance from the Department of Health (DoH) on commissioning arrangements for specialised services was issued in March 2003 and highlights the roles and responsibilities of PCOs to establish effective collaborative commissioning methods¹. It points out that specialised services need to be incorporated in PCOs' future local delivery plans and in Workforce Development Confederations' work programmes, and that decisions of collaborative commissioning groups should be binding on all PCO members. Although mechanisms for commissioning differ in detail in Scotland, Wales and Northern Ireland, these principles are likely to have general application.

The DoH guidance states : "In particular, commissioning groups should ensure that:

- service specifications encompass referral guidelines, access criteria, treatment protocols, standards, NICE appraisal/guidelines;
- there are agreed data sets and a process for monitoring activity, clinical practice and outcomes;
- clinical and financial risk assessments have been carried out for individual specialised services and
- service-specific commissioning consortia and risk-sharing mechanisms are in place where appropriate."

We recognise that these objectives have not all been achieved, but that all involved need to work towards their implementation as soon as possible across the range of conditions. The aim of this document is to catalogue the specialised areas of paediatric care and define their scope, to explain how they link primary, secondary and tertiary care to provide a seamless pathway through care, and to define appropriate patterns of provision. The definitions of conditions needing specialised care are based on those in the second edition Specialised Services National Definitions Set chapter 23 "Specialised Services for Children"². We have also included appendix 1 relating to paediatric dentistry. It is important that those responsible for commissioning health services for children and young people recognise that this list of services does not encompass all those where provision of high quality care locally requires access to specialised advice and expertise. Two examples would be paediatric audiology, where there is increasingly a need for regional networking to deal with specialised services

including cochlear implants, and child protection services where, depending on local factors, a supra-district service may be needed for more complex cases and second opinions.

The level of detail in each section of the report does not imply that these are of equal prevalence, and each section includes information regarding the frequency with which individual conditions are likely to occur. Estimates are based where possible on population prevalence studies. Population prevalence based on birth prevalence may not take into account early deaths and may be an over estimate. For a few conditions actual population prevalence will be less than cumulative incidence due to recovery. **The prevalence and incidence rates given are where possible interpreted as numbers of cases expected in a large PCO with 3,000 live births per annum and 50,000 children under 15 years.** For some very rare conditions expected numbers are given for the population of England or the UK as a whole. This document cannot provide a comprehensive and detailed account of services within each area, but rather aims to give a broad overview of the issues involved when planning services, together with references to more detailed sources. It does not attempt to define the precise workforce requirements for each specialty, though it does indicate the role of skill mix and other innovative solutions, and the extent to which round the clock resident medical cover is needed. It takes account of the views of parents wherever information is available. Further copies are available free of charge from the RCPCH website³. Each specialty group has been encouraged to provide more detailed information on their work on their web site than could be included in this document. Contact details for current representatives of these groups are listed in appendix 2. Local commissioning will also need to take account of factors relating to demography, geography, etc. In order to develop a profile for an individual PCO or group of PCOs it may be helpful to seek advice from and have further discussions with the College representative for that region of the UK. Up to date contact details are available from RCPCH Committee Department (telephone 020 7307 5645 / committees@rcpch.ac.uk). Further information relating to services for children and their families with many of the conditions can also be found through Contact a Family⁴.

Key principles

The Department of Health in England has issued a National Service Framework (NSF) for children⁵. The NSF specifies that children's services should have robust arrangements for timely access to tertiary care when needed, both for emergency transfers to a specialised (regional) centre and for planned referrals. It points out that these arrangements need to cover:

- conditions that are so serious or rare that diagnosis and treatment will be considered specialized;
- severe or intractable cases of otherwise common conditions;
- relatively straight forward procedures but in children with other serious underlying problems;

- procedures that need repeating which were not effective when first performed and;
- simple procedures in neonates and very young patients who need specialised support services such as anaesthetics or neonatal intensive care.

There is an important need for ready access for children with complex disorders to a whole range of services such as genetics, audiology, ophthalmology, specialist nursing, physiotherapy, occupational and speech and language therapy, specialist dietetics, laboratory, pharmacy, radiology, imaging and pathology services which are familiar with paediatric disorders and procedures; also where appropriate palliative care, mental health liaison services, social services and links with education services. In view of the interdependencies of many of the specialised services, there also need to be clearly defined access pathways to paediatric anaesthetic and intensive care services, and the full range of specialists within paediatrics. Supporting services are essential for effective provision of specialised and tertiary services for children, and so must also be appropriately commissioned. All staff need to collaborate closely to ensure that children and their families are receiving consistent and coordinated support, and local commissioning arrangements will need to design services which achieve this. The necessary range of specialised and supporting services can only be sustained at a limited number of tertiary centres in each part of the UK, and high level integration of commissioning will be essential to achieve this.

The NSF also specifies that wherever possible specialised care should be delivered locally in association with other local paediatric services, through outreach services operating within a clinical network. Each network needs to be adequately commissioned, funded and staffed, with clear systems for information sharing, clinical governance, accountability and staff development. Each tertiary service needs to work together with a lead local clinician on behalf of local children's services and primary care to set up referral protocols and arrangements for local service provision. These will include provision of education and training for children and their families so that they can be empowered in their own essential contribution to the management of chronic conditions.

The document further highlights the importance of effective arrangements for transfer of young people from child to adult services, including integration with social care, education and employment. Further information regarding this will be covered in the full NSF when it is published. Local arrangements should include for each specialist condition a policy on timing of transfer, preparation period and education programme, coordinated transfer process, interested and capable adult clinical service, administrative support and primary health care and social care involvement.

Important work has also been taking place in other countries of the United Kingdom. A review of services in Wales has been undertaken, and current work is developing managed clinical networks of care for many of the specialised services for children⁶. In Scotland the National Services Division has been established and has identified standards against which services should be measured⁷.

RCPCH has previously issued two documents of relevance. The first, as the British Paediatric Association in September 1995, provided detailed information and guidance regarding purchase, provision and planning of tertiary services for children and young people. However, the incidence and prevalence of some conditions has changed significantly in the interim, and this will be further affected at local level by population factors such as ethnicity. More recently the College has issued a guide for PCOs on specialist (i.e. paediatric) health services for children and young people⁸. This document should also be consulted as it provides an overview of key elements involved in children's services across the full spectrum. It highlights the importance of resources being available to enable specialist services to comply with national guidelines, the need for accurate data collection, preferably as part of national databases, which are an important component of effective audit and research to improve services for the future. The focus in this present document on specialised and tertiary services needs to be seen in this context of the wider provision of services for children and young people.

In conclusion, I want to thank the authors of each section (Appendix 2), the members of the RCPCH Working Group (Appendix 3), and the many others who worked with us to bring together this document in a short time scale. This would also not have been possible without the expert help and dedication of Krishnan Nayar and Graham Sleight.

John Jenkins

Working Group Chairman

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CARDIOLOGY AND CARDIOTHORACIC SURGERY

1. Definition and scope

Paediatric and congenital heart services are specialised services that are provided by integrated teams of Paediatric Cardiologists and Paediatric Cardiac Surgeons.

1.1 Joint commissioning has generally been undertaken for cardiology and cardiac surgical services. Services are based around supraregional centres which generally offer the following;

- Paediatric cardiac surgery
- Interventional cardiac catheter treatment
- Diagnostic imaging service
- Pacemaker and electrophysiological procedures
- Fetal cardiology service

1.2 Heart and lung transplantation is a national specialist service funded by NSCAG and offered in 2 centres (Great Ormond Street Hospital, London and Freeman Hospital, Newcastle). Similarly ECMO (extracorporeal membrane oxygenation) services for children with cardio-respiratory failure are funded by NSCAG and offered in 3 centres (Great Ormond Street, Freeman and Glenfield Hospital, Leicester)

1.3 Paediatric thoracic surgery for lesions of the trachea, bronchi, lungs and chest wall may be undertaken by Paediatric Cardiothoracic Surgeons. In some centres this work may be undertaken by general Paediatric Surgeons, ENT Surgeons or mainly adult based Thoracic Surgeons.

1.4 Cardiology assessment is also offered in regional hospitals generally by visiting cardiologists from the cardiac centres. Some hospitals also have Paediatricians with an interest in Cardiology, who link in with their local cardiac centre and offer a local diagnostic service.

Important service links:

- Paediatric Intensive Care
- Adult Congenital Heart Disease Service
- General and Subspecialty Paediatrics and Paediatric Surgery
- Neonatology
- Fetal Medicine
- Genetics

2. Incidence and prevalence

Congenital heart disease occurs in approximately 8:1000 live births. 2:1000 will need surgery in infancy and 1:1000 will need surgery from years 2 to 16. Those needing surgery have an

average of 1.6 operations per child. In addition interventional catheter treatment will be required in 2:1000 live births. A typical PCO would therefore expect to have approximately 25 new children with congenital heart disease per year, 10 children requiring cardiac surgery per year and 6 children requiring interventional catheter treatment. Fetal screening identifies 10-20% of children with congenital heart defects and termination of affected fetuses has produced a small drop in the postnatal incidence of heart defects. This has been balanced by the increased prevalence resulting from the improved survival following cardiac surgery in more recent years.

3. Expressed needs

The Kennedy report of the Bristol Royal Infirmary Inquiry included approaching 200 recommendations, many of which have been accepted and enacted. The Department of Health also established a national review group including representatives from cardiac surgery, cardiology, anaesthesia and intensive care, nursing, health service management and parent and patient groups (including the Chair of the Children's Heart Federation, the umbrella group providing national representation of the local parent groups). The Paediatric and Congenital Cardiac Services Review (PCCSR) produced a comprehensive report (www.doh.gov.uk/childcardiac) that was released for consultation in 2001. It focused on service needs, standards and sustainability of the service. The recommendations include many suggestions for improvement including the most radical proposal that surgical centres should have a sufficient workload to maintain high standards (a minimum of 300 paediatric cardiac operations per year) with 3 surgeons per unit (only one hospital currently meets this proposal). If there are to be larger units, this means fewer units and hence political sensitivity. There is also a need to increase the number of paediatric cardiologists. The Minister has accepted the recommendations, apart from that relating to the size of surgical units in the future. Discussions regarding this issue are ongoing, as the status quo is unsustainable. Individual commissioning groups are considering these recommendations in the light of their local situation.

4. Evidence for standards of care

The PCCSR has reviewed the evidence and drawn up a series of standards that can be reviewed on the above website. Each cardiac unit throughout the UK also submits data on surgical and cardiac catheter interventions to the Congenital Cardiac Audit Database (CCAD). This is overseen by a national Project Board under the auspices of the NHSIA, and will be expanded to include fetal diagnosis results from 2004.

5. Current service provision

There are 13 units in the UK providing paediatric cardiac surgery (GOS, Guy's, Royal Brompton, Southampton, Bristol, Oxford, Leicester, Birmingham, Liverpool, Leeds, Newcastle, Glasgow and

Belfast). There are also significant cardiology units in Manchester, Cardiff and Edinburgh that link closely with their nearest surgical centre. Each unit offers a network of outreach clinics for local service delivery when possible.

6. Pathways of care

See www.doh.gov.uk/childcardiac

7. Gaps and pressures

7.1 Training and retaining adequate numbers of Consultant Cardiac Surgeons and Paediatric Cardiologists is challenging, particularly with the need to expand numbers to comply with the European Working Time Directive.

7.2 Recruitment is also challenging for intensive care, theatre, cardiac catheter and cardiac ward nurses, clinical cardiac scientific officers and perfusionists and cardiac radiographers.

7.3 There is an urgent need to establish adequate funding for the development of services for the growing population of adults with congenital heart disease (ACHD). Care arrangements should be seamless and are generally centred around the paediatric unit though some separate ACHD units exist.

COMPLEX CHILD AND ADOLESCENT GYNAECOLOGY

1. Definition and Scope

The majority of this workload is with disorders of sexual differentiation and determination. These are defined as Intersex disorders and related conditions.

Children may present as follows;

- At birth with ambiguous genitalia (congenital adrenal hyperplasia) or associated with other major congenital anomalies (cloacal anomalies).
- During childhood or at puberty with virilisation (5 alpha reductase deficiency and related disorders)
- At adolescence with primary amenorrhoea (complete androgen insensitivity, uterine/vaginal agenesis)
- At adolescence with obstruction of the uterus or vagina (non-intersex).

Clinical input in early childhood is predominantly provided by paediatric endocrinology and urology. Gynaecological input becomes essential prior to puberty and throughout adolescence.

2. Incidence and prevalence

These refer to the estimated numbers of cases throughout the UK.

Condition	Incidence	Female	New Cases per year	Age 0-20yrs requiring follow-up
Congenital Adrenal Hyperplasia (21-hydroxylase)	1:14,199	50-59%	25-30	500
Other forms of CAH	2:1,000,000	Approx 50%	3-5	60
17-hydroxy-steroid dehydrogenase deficiency	1:147,000	Most	3	60
Androgen Insensitivity	1:40,800-1:99,000	Most	4-9	180
Gonadal Dysgenesis True Hermaphroditism	1:100,000	Most	7	140
Mullerian Agenesis/ MRKH	1:5,000 – 1:30,000	All	12	48 (Usually present at approx 16yrs)
Total Cases				988*

(*NB. This total of 988 only represents children raised female and in need of gynaecological input)

3. Expressed Needs

Gynaecological input for these children must ensure and preserve optimum potential for menstruation and future sexual relationships and in some cases fertility. In some conditions surgery may be required prior to puberty to prevent obstruction to menstrual flow. In other conditions treatment can be deferred until late adolescence prior to sexual activity.

The service must be:

Multidisciplinary

Clinical specialities involved include endocrinology, paediatric surgery/ urology and gynaecology. Access to specialist services such as biochemistry and genetics are needed to establish these rare diagnoses. Specialised psychology must be available to families at diagnosis and for ongoing support.

Life long

These conditions are not curable and require lifelong treatment. Adolescence can be particularly difficult for the patient and their family. Psychology services are crucial and seamless transition to a specialist adult service is essential.

Open and Honest

Previous withholding of the diagnosis from patients has led to secrecy and stigma and has deprived patients of access to services. A policy of full disclosure with specialised psychological support is mandatory.

Involve all family members

These conditions are often genetically determined and family members need to access screening and support.

Offer a wide range of treatment for vaginal hypoplasia

There are few long term data for vaginal treatments but poor outcomes from vaginoplasty surgery are reported. Pressure dilation must be available.

Research and Audit

Long term outcome data on all aspects of these conditions is lacking. All patients and families should have the opportunity of involvement in research and service planning.

Involve voluntary groups and service users

Patient groups consulted in this and previous evaluations include:

Androgen Insensitivity Syndrome Support Group (AISSG), Congenital Adrenal Hyperplasia Support Group (CAH), Adrenal Hyperplasia Network (AHN), Rokitansky Support and Advice Group (ROSA)

4. Evidence for Standards of Care

In 1998 a single centre at the Hammersmith hospital was NSCAG designated for gynaecological reconstruction in adolescent and adult women with congenital anomalies of the genital tract. It subsequently became clear that several other units were treating patients as part of multidisciplinary

services for a wider spectrum of conditions. In 2001 NSCAG set up a review group for Intersex. A draft proposal has been prepared and should be available on application to NSCAG. This proposal reviews existing practice and makes recommendations for future service provision for patients of all ages.

5. Current Service Provision

There are currently 14 centres identified in England treating intersex conditions. However the range of services varies and many services are underdeveloped and under resourced. It is accepted that the most comprehensive service is the multidisciplinary joint Intersex service offered by Great Ormond Street Hospital and University College London Hospitals. This service can be used as a model for development of other units.

6. Gaps and Pressures

Psychology services are central to management and are in short supply. Few gynaecologists have the requisite surgical skills although there is a trend away from surgical treatment. Few laboratories can provide full biochemical and genetic investigation.

DERMATOLOGY

1. Definition and scope

The speciality of paediatric dermatology includes a wide range of conditions. There are many severe childhood diseases managed by other specialties which have dermatological manifestations requiring specialist help and it is important that there are close links between dermatologists, paediatricians and many other subspecialties including genetics, plastic surgery & ophthalmology. Most teaching hospitals and many DGHs now have a dermatologist who specialises in paediatric dermatology and network with colleagues.

Table A- range of dermatological conditions

Diagnosis	Incidence	No of active cases per PCT
Inflammatory dermatoses e.g. atopic dermatitis (AD) psoriasis (Ps) Acne vulgaris	AD upto 20% children Ps < 1 % children Affects 80% teenagers	AD - Upto 600 Ps - 30 approx Acne - 2,400 (10-15% need treatment)
urticaria & angio-oedema (Urt)	Very common	Urt - ? 30 - 50
Cutaneous Infections, including warts & molluscum	15% of paed derm referrals	Most children affected by warts at some time
Skin tumours: benign and malignant	20% of paed derm referrals	Melanocytic naevi- almost 100%
Vascular birthmarks: haemangiomas & malformations	Up to 10% children	Up to 300
Hair and Nail disorders	Mild forms common 5% of paed derm referrals	? 30
Pigmentary disorders	uncommon	?
Auto-immune skin diseases	uncommon	?
Genetic skin diseases	Rare but numerous types	?1-5 per annum

2. Incidence and prevalence

There are over 2,000 skin diseases which can affect children (This rises to 3000+ including rare complex genetic diseases which may have dermatological manifestations). Table A shows many of the conditions that fall within this service are sufficiently common to be within the scope of primary care or district based dermatologists. However Table B illustrates conditions which call for more specialised expertise and Table C summarises the services these children may require. Some of these are rare and others difficult cases of common conditions such as eczema or psoriasis.

Table B- Uncommon conditions

Diagnostic group
Epidermolysis Bullosa and other severe blistering disorders
Complex vascular malformations & haemangiomas
Photodermatoses
Giant congenital melanocytic naevi
Severe atopic dermatitis
Other severe inflammatory dermatoses
Severe Cutaneous drug reactions
Cutaneous genetic disorders e.g. the Icthyoses
Difficult vulval problems

Table C – Low volume services provided for large populations

- Phototherapy for severe psoriasis & other inflammatory dermatoses
- Diagnostic Phototesting service
- Specialist paediatric surgery/ plastic surgery and anaesthetics for difficult or malignant tumours and large / difficult vascular problems. Ophthalmological and orthopaedic involvement for some cases
- Specialised radiological imaging such as MRI & MRA for complex vascular lesions
- Laser therapy for vascular lesions. Increasing need for hair removal and pigmented laser treatment
- Dermatoscopy & photography for worrying melanocytic naevi assessment and follow-up
- Specialist dermatological pathology service for difficult skin tumours and dermatoses.
- Team of specialist paediatric & dermatologically trained nurses required for hospital/community – particularly for atopic dermatitis (AD).
- Dietetic support for food allergy/intolerance AD and epidermolysis bullosa. Including inpatient oral food challenge service. Liaison with paediatricians and immunologist for prick testing.
- Genetic counselling and diagnostic services
- Psychological support services required for many severe dermatoses
- Cosmetic camouflage service – Run by the Red Cross

3. Expressed needs

Irrespective of the condition or age of the child, parents tell us that they want timely and accurate diagnosis and assessment, clear honest information and a negotiated and agreed management plan. They require prompt and easy access to specialist nurses to help with therapy which is often very difficult and complicated. Phone-line help is available in some centres. The National Eczema Society (NES) have published a survey on ‘What patients really want’.

4. Evidence for standards of care

The British Association of Dermatologists has published guidelines for many dermatoses and skin tumours including atopic dermatitis (AD), psoriasis, acne, pigmented lesions and phototherapy. These and others are available on the BAD website: www.bad.org There are SIGN guidelines

on the treatment of melanoma. NICE is currently reviewing the use of topical steroids and immunomodulating agents such as tacrolimus and pimecrolimus for atopic dermatitis. Prof Hywel Williams has published a health care needs assessment for dermatology. **Voluntary organisations** include the NES, the Psoriasis association, DEBRA, the Acne Support group, Changing faces, the Red Cross cosmetic camouflage service, the Ichthyosis support group, Alopecia areata self-help group, etc. The British Society for Paediatric dermatology (BSPD) has a web-site: www.bspd.org which links to the BAD and RCPCCH and many of the voluntary organisations

5. Current service provision

Skin disease forms 15% of GP consultations and a high proportion are children. Most GPs have little or no dermatological training. 20% of all UK children have atopic dermatitis but only 10-15% of these are moderate to severe requiring secondary care help. The majority of children referred to secondary care are seen by paediatric dermatologists, or less commonly a general dermatologist –within either the paediatric or dermatology outpatient services. Inpatient services are largely within the paediatric setting except for some older teenagers. Information on service provision is available in a document produced in 2002 by the BSPD.

6. Pathways of care

The majority of children with skin disease could be dealt with at PCO level with some nursing support networking with the secondary dermatology service team. A multidisciplinary approach is necessary for more complex and rare cases and should network across many specialties. Most teaching hospitals can provide the necessary knowledge and skill mix for the majority of difficult cases but this is lacking in some rural areas and extremely rare complex cases may need to be dealt with as a nation-wide service e.g. epidermolysis bullosa, which is commissioned through NSCAG.

7. Gaps and pressures

There is currently no network of paediatrically trained dermatology nurses (particularly necessary for AD) which is urgently required. There is a shortage of dermatologists in the UK & Scotland of around 90-100 posts (6 in Scotland). There are only 4 purely paediatric dermatologists in the UK (3) & Scotland (1) – more posts are required. Specialist networks for managing difficult vascular lesions and other conditions listed in Table B are required in many areas. Pressure from parents to deal with haemangiomas and other vascular lesions is increasing and Laser services will soon be required for most areas These should ideally form part of a vascular lesion care-network rather than being in the private sector. **Emergency and out of hours requirements:** this should be minimal and nurse-run phone help lines can be used

8. Travel

Children may need to visit a distant centre in order to undergo specialist tests or be assessed. Networking and virtual clinics may prevent some of these visits particularly if the team can advise the local hospital about appropriate testing to be done locally.

9. Transition

This is less of a problem in dermatology because children with complex dermatological problems can usually be followed into adult life by their own dermatologist

10. Audit, evaluation and indicators of excellence

There is somewhat patchy registration of rare dermatological diseases at present. However, EB has a national register with DEBRA which is working well and the BAD is attempting to create a register of rare dermatoses. All dermatology departments undergo service audit at local level. There is a published audit on the UK national services for atopic dermatitis. The British Photobiology group has standards of care in place and a programme for ongoing computer audit.

ENDOCRINOLOGY & DIABETES

1. Definition & Scope

Paediatric Endocrinology is concerned with the diagnosis and management of children and adolescents with hormonal disorders (including growth problems). Many specialist paediatricians also care for patients with diabetes mellitus (and unlike adult services paediatric diabetes is never managed exclusively in primary care).

2. Incidence and prevalence

The British Society for Paediatric Endocrinology and Diabetes (BSPED) has recommended that the endocrine problems in Table A are suitable for management within the scope of a district or PCO based team with appropriate training (and where necessary, appropriate support).

For those conditions where active cases are highlighted (*) these numbers are potential cases, based on auxological cut-offs; i.e. for stature: height <2nd & >98th centile, for weight: BMI >+2SDS, for puberty: percentage of Caucasian girls showing breast development by 8 years of age.

	Incidence	No of active cases per PCT
Short & tall stature	1 in 40	1,250*
Delayed puberty	1 in 30	1,000*
Obesity	1 in 50	1,250*
Idiopathic central precocious puberty (girls)	1 in 20	150*
Congenital hypothyroidism	1 in 4000	12
Idiopathic growth hormone insufficiency	1 in 3000	16
Diabetes mellitus	1 in 400	125

Table A – range of conditions

Table B illustrates those rare conditions that call for more specialised expertise. These are usually managed in tertiary centres and Table C summarises the additional services that these children may require. There are currently no quaternary services (e.g.. NSCAG) funded in paediatric endocrinology & diabetes, although some are being developed (e.g.. PHHI (persistent hyperinsulinaemic hypoglycaemia of infancy)).

Table B – Uncommon conditions

ENDOCRINOLOGY

Adrenal disorders (incl CAH)
Thyrotoxicosis
Intersex disorders
Turner syndrome
Hypopituitarism
Growth & pubertal problems in chronic renal failure
Care of endocrine problems in cancer survivors.
Rare endocrine tumours
Disorders of calcium metabolism
Hypoglycaemia

DIABETES:

Diabetic complications
Secondary, type 2 or rare forms of diabetes (eg. DIDMOAD)

Table C – low volume services provided for large populations

- Specialist nursing staff for endocrine testing and also training of patients & staff.
- Specialist psychological support to affected patients and their families.
- Combined multi-disciplinary endocrine clinics with other specialties eg. Intersex disorders (in association with CAHMS & child/adolescent gynaecology), Oncology, Turner & Prader-Willi syndrome.
- Provision (where appropriate) of outreach endocrine services to local units.
- Daytime and on-call provision of support in diagnosis and advice on management of uncommon conditions.

3. Expressed needs

BSPED & its members work very closely with appropriate family support groups (notably the Child Growth Foundation (CGF), Turner Syndrome Society (TSS) and Diabetes UK). Specific information for families and their children has been produced, often written by both support groups and paediatricians. This is available on websites (including the BSPED), through family support groups themselves and through departments of endocrinology & diabetes.

4. Standards of care

Recent NSFs in Diabetes <http://www.doh.gov.uk/nsf/diabetes> and Children (standards) <http://www.doh.gov.uk/nsf/children.htm> have highlighted the specific needs of patients with diabetes and chronic disorders (including endocrine problems). The management of other specific groups (e.g.. GH therapy, Insulin pumps) have been dealt within the remit of NICE <http://www.nice.org.uk>. Consensus evidence-based guidelines are also being established for a number of conditions (e.g.. GH-insufficiency, CAH, transitional care), both nationally (BSPED) & internationally (ESPE). A national shared care protocol is currently available for GH therapy, and a number of other proto-

cols are in the process of being written e.g. GnRH agonists in central precocious puberty.

5. Current service provision

Previous national & international guidelines have been produced for paediatric diabetes. The diabetes NSF has highlighted the need for specialist teams to work with children with diabetes, although repeated national audits performed over the last decade continue to demonstrate that few units currently provide these facilities.

Historically tertiary endocrine units have been based at the growth centres (N=25) which were established several decades ago, with a specialist paediatric endocrinologist/diabetologist. These are now increasing both in size and number, as are the number of linked subsidiary secondary units (general paediatricians with an “interest”) who provide local diabetes care and secondary endocrine care (which may be supported by an outreach service).

Detailed workforce information in paediatric endocrinology & diabetes is currently available on the following websites: BSPED (www.bsped.org.uk) and RCPCH (www.rcpch.ac.uk).

6. Pathways of care

The referral pathways (especially for growth disorders) are being reviewed in the light of *Health for all Children* (4th Edition) which does not recommend routine screening for children until school entry. The BSPED is currently putting together prospective studies to obtain an evidence base for screening. In addition to secondary and tertiary referrals from primary and secondary care, paediatric endocrinologists are increasingly being referred patients with complications of other chronic paediatric diseases (e.g. cancer, renal, gastro-intestinal, respiratory, rheumatology).

7. Gaps in Service

As with other paediatric specialties paediatric endocrinology is suffering from a lack of suitable trainees (only 4 of 8 NTN posts filled last year). There are also insufficient numbers of appropriately trained nurses, dieticians and psychologists. Increasing numbers of patients (e.g. diabetes (type 1 & type 2), obesity and chronic illnesses) are putting pressure on the service, as many require long-term follow-up.

In addition, many of these patients (all diabetics, and many endocrine disorders) require transfer to adult services (preferably via an adolescent service and hand-over clinic), and usually once growth has been completed. Reassessment may also be required at this stage. Given that a number of patients are managed in stand-alone children’s hospitals, this process is often not seamless.

8. Audit, evaluation and indicators of excellence

A national register for patients with diabetes mellitus is already in place through Diabetes UK, and as part of the NICE process it has been agreed that there will be a national register of new patients commencing GH therapy. There is in addition a near-national system for safety and efficacy monitoring of GH (KIGS). This, and other audits of endocrine management, are being organised through the BSPED (including the Clinical Trials Unit). Management protocols of a number of diseases (e.g.. GHI, CAH, transitional care) are being coordinated internationally in association with the European umbrella group (ESPE).

GASTROENTEROLOGY, HEPATOLOGY, AND NUTRITION

1. Definition and scope

Paediatric gastroenterology is a clinical speciality comprising the investigation and management of disorders of the gastrointestinal tract (the oesophagus, stomach, pancreas, small intestine and colon) in infants and children. It also encompasses two related specialities: first, paediatric hepatology (liver diseases – see below) and second, clinical nutrition; the latter because diseases of the gastrointestinal tract are potent causes of nutritional disorders, often requiring specialised nutritional care. Whilst a proportion of paediatric gastroenterology and nutrition belongs within primary and secondary care there are elements which by virtue of their rarity in general paediatric practice, the severity of the condition (e.g. severe constipation or gastro-oesophageal reflux), the specific difficulties encountered in very young children or the complexity of their management (e.g. intestinal failure) means that they must be considered by tertiary or specialist services.

The aspects of paediatric gastroenterology that should be regarded as specialized are:

Gastroenterology

- Intestinal failure
- Inflammatory bowel disease
- Gastrointestinal bleeding
- Complex motility disorders including the severe end of constipation and gastro-oesophageal reflux
- Protracted diarrhoea (greater than 3 weeks duration and weight loss)
- Congenital transport disorders
- Multiple food intolerances
- Coeliac disease (unless pathology and paediatric dietetic service available locally)
- Pancreatitis
- Pancreatic exocrine insufficiency other than cystic fibrosis

Nutrition

- Home parenteral nutrition
- Nutrition for complex disorders whether supportive or primary treatment
- Gastrostomy and jejunostomy tube feeding

Hepatology

- Neonatal hepatitis syndromes
- Childhood chronic liver disease
- Surgical liver disease

- Jaundice not due to uncomplicated hepatitis A infection

Specialised investigations

- Upper and lower GI endoscopy
- Liver biopsy
- Intestinal biopsy
- Rectal biopsy
- Oesophageal pH monitoring
- Breath hydrogen tests
- Pancreatic function tests
- Motility studies and manometry

Supra regional services in paediatric Hepatology

There is a national service for paediatric hepatology funded by the National Specialist Commissioning Advisory Group (NSCAG) and provided in 3 national units. This service covers liver failure, paediatric liver transplantation, surgery for biliary atresia and paediatric intestinal transplantation. There are established shared care arrangements and referral pathways between the three national units and specialist gastroenterology units. The three supra-regional units at King's College Hospital, London and The Children's Hospital, Birmingham and St James's University Hospital, Leeds. Refer to guidelines for services provided by paediatric hepatology. Children's NSF document at <http://www.doh.gov.uk/nsf/children.htm>

Paediatric gastroenterology is an essential part of a region's tertiary service profile. The service has important interfaces with a number of related medical disciplines:

- general and neonatal paediatrics
- neonatal, paediatric and adult surgery
- oncology
- inborn errors of metabolism
- immunology and infectious diseases
- gastroenterology and hepatology in adults

2. Incidence and Prevalence

Paediatric gastrointestinal disorders include a wide range of diseases. The incidence and prevalence data is difficult to ascertain, for example constipation and gastro-oesophageal reflux are common disorders most of which are managed in primary or secondary care, with only difficult cases being referred for specialist input. Specialist units do however have an obligation to be part of the process of setting up shared care guidelines and indications for referral. They need to be able to see and promptly assess difficult cases as part of that shared care.

Inflammatory Bowel disease Childhood Inflammatory bowel disease is much less common and is within the National Specialist Services definition set, and as such should have management of all cases led by specialist centres. It represents a significant part of a paediatric gastroenterologist's workload, and there is good epidemiological data available. Childhood IBD has a prevalence of 20 cases per 100,000 children under age 16 years, with an incidence of 5 new cases per 100,000 children per year. This means a PCO with 50,000 children will have 10–12 children with inflammatory bowel disease. This does not take into account children/adolescents between ages 16 and 18 many of whom are managed in paediatric or transition clinics with paediatric input. These estimates are likely to be significantly underestimating the scale of the problem, as many centres are seeing between 20-50 new cases per year. This disease has a chronic relapsing course with a high morbidity and need for specialist input and investigation. All children with inflammatory bowel disease should therefore have access to a specialist centre for assessment and management.

There has (nationally) been an increase in the incidence in inflammatory bowel disease in childhood. Inflammatory bowel disease is designated as a specialist service in the National Specialised Services Definitions Set prepared by the London Regional Specialised Commissioning group. The British Society of Paediatric Gastroenterology, Hepatology and Nutrition recommend that all children with inflammatory bowel disease be managed jointly with a Paediatric Gastroenterologist.

Endoscopy. Paediatric endoscopy is increasingly centralised in specialist centres. Many factors have influenced this change and the increase in demand for paediatric endoscopy. On a survey of most units in the United Kingdom carried out by the BSPGHAN in 1999 the clinical activity (which is usually an underestimate of clinical need due to constraints of resources and manpower) revealed the following (figures are expressed as numbers of procedures carried out per annum in the age group under 16 years but the denominator is per 100,000 of the *total* population of all ages): upper endoscopy 20.5/100,000; ileo-colonoscopy 6.4/100,000. It is the opinion of the Council of the BSPGHAN that these figures required updating and hence a survey was carried out in 2003. The corresponding figures revealed: upper endoscopy 23/100,000 and colonoscopy 7.5/100,000.

Change in clinical practice has occurred with an increased need for endoscopy which is evidence based – increased yield of upper GI endoscopy in Crohn's disease, increased need for upper GI endoscopy in children with reflux to exclude allergic oesophagitis, need for children who previously had sigmoidoscopy to have ileo-colonoscopy:

- Change in practice with virtual cessation of Crosby capsule biopsies to confirm the diagnosis in Coeliac disease and replacement by endoscopic biopsies
- Increased prevalence of Coeliac disease from 1 in 2000 to 1 in 2-300

- Increased referrals for biopsy confirmation of Coeliac disease
- Change in practice with increased use of General Anaesthesia for endoscopy
- Centralisation of service in line with patient safety, clinical governance and national recommendations with therefore the need to provide a regional rather than district service for endoscopy in children

3. Expressed needs

Children and young adolescents with paediatric gastrointestinal or nutritional problems – known or suspected - should have rapid access to specialist input as part of a managed clinical network. Other tertiary services (e.g. intensive care, cardiology) should be able to access paediatric gastroenterology easily and preferably on the same site, in order that children with multiple problems are managed in a child centred way with access to all their needs in one setting. This should enhance dialogue between the often multiple clinicians involved in the care of a child with complex problems. This means a paediatric gastroenterology unit as configured below and practising in the way described, should be allied to every centre seeing children with specialist health problems, and should be available and accessible for children from the local and regional PCOs.

We have worked in partnership with our parent and child support organisations in providing this recommendation

Crohn's in Childhood Research Appeal (CICRA) <http://www.cicra.org/>

National Association for Colitis and Crohn's Disease (NACC) <http://www.nacc.org.uk/>

Children's Liver Disease Foundation (CLDF) <http://www.childliverdisease.org/>

Patients on Intravenous and Nasogastric Nutrition Therapy (PINNT) <http://www.pinnt.com/>

Coeliac society <http://www.coeliac.co.uk/>

Cystic fibrosis trust <http://www.cftrust.org.uk/site/>

4. Evidence for standards of care

There is guidance on the management of children provided through the Guidelines for Purchasers of Paediatric Gastroenterology, Hepatology and Nutrition. There are few evidence based protocols available, but there is evidence that emphasises examples of patchy quality of care outside specialist centres. This is particularly the case for children with inflammatory bowel disease, with failure of jejunal biopsy sampling and the inappropriate use of gluten-free diets in children suspected of having coeliac disease. Furthermore, outcomes are improved for children receiving parenteral nutrition in a specialist centre with a multidisciplinary nutritional care team. There is also guidance available through the British Society of Paediatric Gastroenterology, Hepatology and Nutrition,, British Society of Enteral and Parenteral Nutrition, British Society of Gastroenterology, European Society of Paediatric Gastroenterology, Hepatology and Nutrition and North

American Society of Paediatric Gastroenterology, Hepatology and Nutrition. NICE guidelines have been produced for gastrointestinal problems but not specifically for children.

5. Current service provision

Within the United Kingdom, the management of children with paediatric gastrointestinal disorders remains patchy, without patient focused organised clinical networks or pathways. Fundamental to such models are the paediatric gastroenterologist, general paediatrician and general practitioner, supported by other medical and allied professionals. There are currently less than 20 paediatric gastroenterology units allied to teaching centres, mostly staffed by between 0.5 and 1 WTE consultant paediatric gastroenterologists, although some with 3. In order to provide services as per the national service framework these centres will need considerable expansion in consultant staff.

Progress is being made with specialist centres providing outreach clinics and acknowledging the importance of the general paediatrician with an interest in paediatric gastroenterology in a district hospital. It is essential that the latter has access to the facilities and support services available in the tertiary centre, and is part of a unified service for children which is appropriate for the area they serve

6. Pathways of care

Paediatric Gastroenterology should be provided as part of a managed clinical network. Each specialist centre should aim to provide the above services to referral centres. The child needs to be at the centre of such a model. Centralised expertise should be easily accessible.

The following would be essential for this system to function:

- Access to expert opinion by telephone 24 hours/day.
- Ability for rapid outpatient referral to defined 'urgent / emergency referral clinic'.
- Capacity to accept in-patient transfers at short notice.
- Capacity to admit children directly for specialist investigations without prior clinical assessment.
- Regular joint outreach clinics at referring hospitals to assess new patients and review shared care patients as necessary.
- Designated paediatrician at shared care hospital with adequate support services to provide shared care and act as referring consultant to specialist centre.
- Rapid communication of out-patient and in-patient management plans to designated referring consultant.
- Regular meetings between teams within managed clinical network to review guidelines / communication / training needs.

7. Gaps and pressures

Patchy service provision exists throughout the country with, using present WTE consultant staff as a marker, single-handed specialists serving large regions. This has significant impact on access to services, development of managed clinical networks including outreach, out of hours cover, training, research and audit. The European Working Time Directive has further exacerbated this with the reduction in junior doctor availability to support the service. This is leading to a fundamental change in how health care is managed with a switch from consultant led to consultant delivered service. This will need to be addressed by the appointment of more consultants in regions where there is a deficit. The appointees need to fulfil the training requirements as set out by the CSAC. It is essential however to ensure that adequate support services accompany this expansion in order to ensure the complete service is offered. This should include funded endoscopy lists, access to specialist radiology and histology services, as well as dietetic and nurse specialist support. This will be necessary to ensure easily accessible, safe and effective care, which is up to date and can provide 24 hour access for the vulnerable group of children and adolescents with paediatric gastroenterology and nutritional disorders.

The requirements for a specialist unit are:

A population and referral base of sufficient size (approx. 2 million) to justify the appointment of 3 gastroenterologists to provide on-call cover. Exceptions should be made for remote areas.

Trained paediatric gastroenterologists who fulfil the RCPCH/BSPGHAN higher professional training criteria and who have accreditation with sub-specialist recognition on CCST.

An endoscopy service, provided in a child-friendly setting, with at least 75 procedures per consultant endoscopist per annum.

Provision of anaesthetic support for paediatric endoscopy.

Specialist nursing staff, with particular training in nutritional care and paediatric intensive care / recovery (for management of children having procedures under sedation / GA).

Paediatric gastroenterology clinical nurse specialist.

Easy access to the full range of specialist services for children on the same site, including specialist paediatric/adult surgery and intensive care. Similarly, specialist centres undertaking complex neonatal and paediatric gastrointestinal surgery should have access to a full range of gastroenterological and nutritional support services.

A full range of diagnostic/endoscopic services, including:

Radiologists with specific training in paediatrics and expertise in paediatric diagnostic procedures

Histopathologists with expertise in paediatric gastroenterological pathology

oesophageal pH monitoring

pancreatic function testing*

small intestinal disaccharidase assays*

Breath testing

Motility studies

Liver Biopsy and ERCP*

**some centres only*

Paediatric dietetic services, staffed by dieticians who spend the majority of their time working with children and available to provide input to the wards on a daily basis and to work alongside the clinicians supporting gastroenterology clinics and gastroenterology and nutrition ward rounds

Paediatric pharmacist

Psychological support from within child psychology / psychiatric services

Parent's accommodation and support services catering for the special needs of children including social workers, play therapists and teachers

A multi-disciplinary nutritional care team which will include a paediatric speech and language therapist for assessment of children with feeding difficulties

Close links with adult gastroenterologists and an established mechanism for handing on adolescent patients to gastroenterological services for adults

A commitment to undertake outreach clinics and shared care with general paediatrics

References

Children's National Service Framework <http://www.doh.gov.uk/nsf/children.htm>

National Specialised Services Definition Set <http://www.doh.gov.uk/specialisedservicesdefinitions/23children.pdf>

DGH GI, Hepatology and Nutrition Services: *Final Report* <http://bspghan.org.uk/document/DGH%20SERVICES%20BSPGHAN.PDF>

Guideline for Purchasers of Paediatric Gastroenterology, Hepatology and Nutrition <http://bspghan.org.uk/document/GuidelinesforPurchasersSeptember2003.doc>

IMMUNOLOGICAL DISORDERS/INFECTIOUS DISEASES/ALLERGY

Infectious Disease

1. Definition & Scope

(Specialised Services National Definition Set: Children - Definition No 23 [section 5], Infectious Diseases [adult] - Definition No 18).

Much of the work within this speciality is consultative; including prevention of infection; treatment of infections occurring in children treated by other specialists; and liaison with laboratory and public health specialists. There is also coordinated work with adult teams (e.g. for Tuberculosis (TB) or prevention of perinatal transmission of HIV, Hepatitis).

The following conditions are managed within the remit of Paediatric Infectious Diseases:

- Severe paediatric infections
- Rare paediatric infections
- Inflammatory conditions affecting children
- Neonatal Infections and congenital infections
- Blood borne infections and sexually transmitted infections
- Infections in the immunocompromised
- Imported infections
- Worldwide emerging new infections
- Prevention of paediatric infections
- Hospital -acquired infection /infection control / antimicrobial resistance
- Specialist knowledge of antimicrobial treatments for children

2. Incidence and prevalence

National standardised incidence and prevalence rates for a typical PCO cannot easily be applied to paediatric infections as these are heavily influenced by ethnicity; migration; dispersal; and local factors. In general the burden of paediatric infection is greater in urban areas. Local public health planning within PCOs is therefore very important. National surveillance of a number of important infections is currently undertaken, including: HIV; Hepatitis; TB; Sexually Transmitted Infections; Meningo-encephalitis; Imported Infections etc.

3. Expressed needs

The need for improved co-ordination and provision of services for control and treatment of

infections in adults and children has recently been delineated: “Getting Ahead of the Curve” - A strategy for infectious diseases www.doh.gov.uk/cmo/idstrategy; Developing Infection Services for Children RCPCH, BPAIIG, draft 160902; House of Lords report on developing services for infection in England 2003 www.parliament.uk/hlscience; London HIV Consortium Paediatric Sub-Group - Developing Service Networks in Paediatric HIV Infection 2003; a report is in preparation from the London paediatric TB working group.

4. Key voluntary organisations

HIV organisations; Parliamentary HIV Group; Refugee health Groups; Kawasaki Disease Society; Meningitis Research. Infectious disease in children is often concurrent with poverty, mobility, language and cultural barriers and these disenfranchised families are often not well represented in either the voluntary or statutory sectors.

5. Evidence for standards of care

HIV treatment BHIVA + PENTA guidelines available via CHIVA (www.bhiva.org/chiva/index.html)

Richardson M et al. Evidence level of incubation periods; periods of infectiveness and exclusion periods of communicable diseases for schools and preschools. *Pediatr Inf Dis J* 2001;20:380-91.

Brogan PA et al. Kawasaki disease: an evidence based approach to diagnosis, treatment, and proposals for future research. *Arch Dis Child*. 2002 Apr;86(4):286-90.

Pollard AJ et al. Emergency management of meningococcal disease. *Arch Dis Child*. 1999 Mar;80(3):290-6.

RCPCH. Working Party Report. Immunisation of the Immunocompromised Child; Best Practice Statement. 2002 at www.rcpch.ac.uk

NICE TB treatment guidelines currently in development

BTS TB guidelines.

A national evidence based protocol for antibiotic prescribing in common infections of childhood is currently being written by members of the BPAIIG.

Winning Ways: Working Together to Reduce Healthcare Associated Infection in England - www.doh.gov.uk/cmo/hai/winningways.pdf

6. Current service provision

Before 1980 most children with infection were treated by adult ID physicians or general paediatricians. Since then services have developed in a small number of centres due to academic interest, existing adult ID services; and paediatric immunodeficiency services. There has been no

central planning and so provision is patchy depending on academic, immunology or HIV services. This is in marked contrast to the advanced development of the service in North America / Europe (www.pidsa.org, www.espid.org). Provision of specialist nurses, pharmacists, and other multi-disciplinary team members also remains inadequate in most places. To improve access to optimal care for all children with rare / common infections a service network for paediatric ID has been proposed, similar to that already in existence for paediatric oncology (see below). Such a shared care network is already well underway for children with HIV (see HIV section), but remains in its infancy for other infections.

7. Pathways of care

The proposed service network has a regional “Infection Centre” supporting more local “Infection Units”. An Infection Centre would: be linked to a medical school; have clinical inpatient ID beds for children and adults with specialised facilities e.g. negative pressure isolation for infections such as multi-drug resistant TB (MDRTB); have links to other specialties; be active in multidisciplinary training and recruitment; lead continued professional development; and lead regional data collection, audit, research and development. Infection Units would be set up in district general hospitals with a lead clinician (paediatrician / nurse specialist) working within the local infection team. Children with more complex / rare problems would have shared care with the Infection Centre where necessary.

For infections affecting adults and children (e.g. HIV, hepatitis, TB), family clinics are needed. Adherence support nurses and specialist pharmacists are vital to reduce the risk of development of resistance to long-term treatments e.g. for HIV / TB. Involvement of local user groups in setting up clinics helps to strengthen individual responsibility for health as well as better understanding between patients and providers. Transitional adolescent clinics for young people with HIV moving to adult care have been established in a few centres, but should become the norm for this condition. Appropriate STI services should also be provided for adolescents. Supra-regional responsibility for certain rare and complex infectious problems should be considered (e.g. for children with viral haemorrhagic fevers.)

Such a network made up of nationally identified teams would undoubtedly improve care and access to care for children with infections across the UK and ideally should be defined within the NSF for children.

8. Gaps and pressures

The proposed national service network for paediatric ID requires local planning, identifying and training the paediatric ID unit lead in each DGH, and the appointment of paediatric infectious diseases specialists in tertiary paediatric centres in the UK. Investment in new posts will be

required to achieve this.

Training of paediatric ID specialists according to the syllabus approved by the BPAIIG and RCPCH specialist CSAC is undertaken in a few accredited centres and posts are allocated by national competition with only 3 places per year. Allocation of training numbers will need to be reconsidered to cater for the unmet need nationwide. Training courses for paediatric specialist nurses also need to be set up.

Allergy

1. Definition and scope

This specialty includes the full range of atopic disease as described in Specialised Services for Allergy (all ages) - Definition set no. 17.

Links with other specialties include: gastroenterology/nutritional support, respiratory paediatrics, paediatric intensive care, paediatric A&E, immunology, dermatology, paediatric ophthalmology, child psychiatry and paediatric ENT surgery.

2. Prevalence

	Prevalence	No. of active cases per PCT
Atopic dermatitis	22 %	11 000
Asthma and wheezing	21 %	10 500
Allergic rhinoconjunctivitis	10 % (33 % of 13-14 yr olds)	5 000 (1,980 13-14 year olds)
Food allergy	8 % (20 % of 2-3 year olds)	4 000 (1,200 2 - 3 yr olds)
Multiple food allergies	0.5%	250
Children at risk of anaphylaxis	2%	1 000

Allergic disease comprises a significant percentage of the workload of primary care. More rare conditions, or conditions calling for more specialised expertise should be managed in a specialist paediatric allergy centre, including anaphylaxis, angioedema, chronic urticaria, severe and multi system allergic disease, multiple or severe food allergy, drug allergy, venom allergy, latex allergy, vaccine reactions, where the diagnosis of allergy is in doubt (for example discordance between history and testing), and excluding allergy as a cause of disease or abnormal behaviour (ADHD affects 10-15% of children; 7 500 children per PCO). These conditions often co-exist in atopic children.

Services offered by specialist paediatric allergy centres include:, a full range of diagnostic testing, day case challenge testing, immunotherapy facilities (in accordance with national guidelines), a full time advisory service including allergen avoidance and primary and secondary allergy prevention, co-ordination with a community paediatric team for management of children at risk of anaphylaxis in nurseries and schools, specialist dietetic service, education, teaching and providing a resource

for GP's and other consultants throughout the region

3. Expressed needs

Documents highlighting the need for further provision of allergy services include:-

1. Allergy the unmet need - a blueprint for better patient care. Royal College of Physicians 2003
2. Stolen Lives; The allergy report-the impact of allergies on peoples lives in the UK today. Allergy UK 2003
3. Bridging the Gaps - commissioning and delivering high quality integrated healthcare. The Respiratory Alliance 2003

What patients need

Anxiety and lack of information among families affected by allergies cause distress and patients commonly report that doctors give inadequate help and information. They need appropriate management, education in allergen avoidance and the use of inhaler devices.

Key voluntary organisations

Voluntary organisations contacted include: The Anaphylaxis Campaign, Allergy UK, National Asthma Campaign and British Lung Foundation.

4. Evidence for standards of care

Standards of care are documented in the Royal College of Physicians document '**Good Allergy Practice**' 1994 and '**Allergy: the unmet need**', Royal College of Physicians (2003). It describes current deficits in NHS allergy services and recommends the development of a coordinated and integrated allergy service over the next 10 years.

5. Current service provision

Children with allergies are managed by general paediatricians or organ based specialists (paediatric gastroenterologists, respiratory paediatricians), ENT surgeons and dermatologists. Whilst these specialists have an important role in the management of allergic disorders, a partnership needs to be developed with specialists in paediatric allergy. Many children are seen in appropriate adult allergy clinics. Other children are dealt with by their GP's, who have no clinical training in allergy. In regions with a non-existent services (much of the UK) allergy lacks a voice. Allergy is often confused with immunology.

6. Pathways of care

Allergy needs a 'whole system' approach in which it is treated as a condition in its own right rather than as a series of diseases depending on the organ system involved. Most patients with simple allergic disease will be dealt with in general practice. It is envisaged that allergy services will progressively become primary care led, with expertise from the hospital setting for more severe and complex problems.

At PCO level, children with allergies could be managed by a team comprising the general practitioner, practice nurse, the practice lead in allergy and GP's with a special interest in allergy offering allergy diagnosis and testing, symptom management and referral to specialist services where appropriate.

General paediatricians with an interest in allergy in district general hospitals should deal with local needs. One model may be for a shared appointment between trusts and a regional allergy centre. A designated community paediatrician within each primary care trust would co-ordinate management. General paediatricians networking with specialist centres will continue to have primary responsibility for patients with single organ or uncomplicated allergic disease.

'Allergy; the unmet need' proposes the development of regional allergy centres to manage more complex cases, offering equality of access throughout the UK. Each region should have a specialist allergy centre staffed adequately with consultants in paediatric allergy supported by paediatric nurse specialists and paediatric dieticians with facilities for training SpR's in paediatric allergy and general paediatricians with an interest in allergy. Regional allergy centres will provide specialist expertise for managing difficult allergic disease throughout their region (tertiary care), care for allergic disease in the local population which cannot be dealt with in general practice (secondary care), act as an educational resource for the region, network with and enable local training in allergy for general paediatricians, support training at local level for general practitioners and nurses in the management of common allergies in primary care and to be supported by appropriate laboratory resources for in vitro allergy testing.

7. Gaps and pressures

Given the increase in allergy referrals, primary care must provide front line care for allergy, with support from paediatric allergy specialists working in hospitals. Specialists in paediatric allergy, working in regional allergy centres, are needed to achieve and maintain appropriate standards of care. There is a need for facilities for accurate diagnosis and management of paediatric allergies, day case facilities for challenge testing and allergen immunotherapy in appropriate settings. New and expensive ways of treating common conditions, such as the use of anti-IgE to treat food

allergy, will need careful assessment and supervision. Clinical leadership must initially come from specialist centres diagnosing and managing the most complex cases and supporting the development of capacity within primary care.

Paediatric Immunology

1. Definition and Scope

Paediatric immunology includes the investigation, clinical assessment and management of patients with primary immune deficiency, complex disorders of immuno-regulation as well as input into the management of patients with severe allergy, unusual infections and systemic auto-immune disease. There are also important links with paediatric respiratory medicine, gastroenterology and haematology / oncology.

2. Incidence and Prevalence

Severe primary immune deficiencies are rare disorders, but in a region with a total population of two to three million, there are approximately 200 referrals per annum and 40 children on intravenous immunoglobulin replacement therapy. Services offered include investigation and delineation of primary immune deficiency disease, referral for bone marrow transplantation in a supra regional centre for children with one of the more severe forms of primary immune deficiency, initiation of intravenous immunoglobulin therapy for antibody deficient patients, and the management of complications (such as bronchiectasis and gastrointestinal disease) and unusual infections in immunodeficient patients. Other roles include supervision of immunomodulation with high dose immunoglobulin therapy, monoclonal antibodies to cytokines and cytokine receptors in severe and unusual inflammatory disease, and the assessment of immune function in patients with secondary immune deficiency (e.g. from immunosuppressive therapy).

3. Expressed Need and Evidence for Standards of Care

Consensus document for the diagnosis and management of patients with primary antibody deficiencies, Royal College of Pathologists, 1995.

Families are increasingly concerned about the possibility of immune disorder. At the same time knowledge has increased exponentially and there is a real need for specialist paediatricians who have the knowledge and expertise to properly diagnose immune deficiencies when they do exist, and to be able to reassure patients and families when they don't exist. The outcome from bone marrow transplantation has improved greatly, but this is still dependent on early and rapid diagnosis. Ongoing supportive treatment, for example in antibody deficiency or in Chronic Granulomatous Disease, is also more effective, but again is dependent on early diagnosis, proper monitoring and supervision of ongoing treatment by a fully trained and resourced multi-disciplinary team. Patients therefore need a regional centre where a paediatric immunologist works alongside paediatric

infectious disease and allergy specialists, as well as the organ sub-specialists, and provides a mainly out-patient and consultative service. There is also a need for specialist input from a paediatric immunology nurse, and it is vital that there is the backup from a fully accredited immunopathology laboratory with a consultant immunologist / immunopathologist in charge.

Key voluntary organisation: the Primary Immunodeficiency Association (PiA).

4. Current Service Provision

NSCAG (National Specialist Commissioning Advisory Group) funds two supra regional centres for the assessment and treatment of children with the most severe forms of primary immune deficiency.

Service provision is 'patchy' and in many regions there is no paediatric immunologist at all. In some centres children are seen by adult clinical immunologists, in others by an interested paediatrician from an associated sub-specialty (for example: paediatric respiratory). There is widespread recognition that this is not satisfactory, and that in this rapidly changing field where outcomes are improving so much, it is vital that properly trained sub-specialists' advice is available in each region.

5. Pathways of Care

Paediatric immunology needs a network approach so that standards of care can be maintained and difficult cases discussed appropriately.

The major paediatric immunology centres currently hold outreach clinics, for example: the 'Northern Network' includes Edinburgh, Newcastle, Middlesbrough, Carlisle, Manchester and Dublin, with less formal, but nevertheless effective links that include Liverpool, Sheffield, Leeds, Hull and Nottingham. Joint clinics between paediatric respiratory physicians and rheumatologists are also useful. Within each centre one would expect the paediatric immunologist to work closely with paediatric infectious disease physicians and paediatric haematology / oncology specialists, providing cross over cover for leave etc.

6. Gaps and Pressures

Most regional centres do not have paediatric immunology and there is an urgent need for this. Such specialists can't exist single handed, but as a team with other sub-specialists and adult clinical immunologists / immunopathologists, can then form part of a network with the supra regional centre.

HIV Infection

1. Definition and Scope

This specialty includes the long term care of children born to HIV infected mothers and horizontally infected with HIV.

Links with other specialties including Adult ID, GUM, and paediatric infectious diseases.

2. Incidence and prevalence

In the UK there is a higher incidence of maternal HIV infection in London and the South East. This is changing rapidly with increasing dispersal of asylum seeking families across the UK. Currently in England around 700 confirmed pregnancies to HIV positive mothers are reported nationally through the RCOG each year. Taking account of additional reports there are now around 1000 infants /year born in the UK to HIV infected mothers. All these infants will now need long term follow up. The uptake of widespread antenatal HIV testing, treatment in pregnancy with antiretroviral therapy, Caesarian section and bottle feeding has led to a dramatic reduction in vertical transmission to around 1-2%. Most of these infants have been exposed to highly active antiretroviral therapy (HAART) in utero, and the long term safety is unknown. 1058 HIV infected children had been born in the UK by end June 2003, of who 224 are known to have died. Around two thirds live in London and the South East. Most new children being seen in the UK are now born abroad, and the rate of new diagnoses has not decreased.

3. Expressed needs

The dramatic reduction in mortality associated with the introduction of HAART has been documented (CHIPS - BMJ 2003). In a cohort study of over 600 HIV infected children, the median age is 7.8 years. Mortality has fallen by nearly 90% since 1996, with admission rates also falling from 4.3/100 child years of follow up in 1996 to 0.7 in 2001. Nearly 75% of children under follow up are on HAART. This is very complex, expensive treatment with many side effects. Around 20 different drugs are currently used. All prescribing should only be done in conjunction with a paediatric HIV specialist. Families need high quality local service provision with a multidisciplinary team. They also need HIV specialist voluntary and state support services.

Key voluntary organisations

Terence Higgins Trust; Avert; Body and Soul; Positively Women; Body Positive; African HIV Forum

4. Evidence of Standards of Care

European guidelines for the management of children with HIV are published (www.ctu.mrc.ac.uk/penta). The UK fully endorses and uses these guidelines. The Children's HIV Association provides guidelines for the management and treatment of all aspects of paediatric HIV infection in the UK (www.chiva.org).

5. Current service provision

In London the care of children with HIV developed at the 3 Paediatric Infectious Diseases Units in London (GOS, St Mary's and St George's). The model of care - the Family HIV Clinic where adults and children can be seen together - has been established for over 10 years in each of these centres. Other centres have now developed local care provision. The London HIV Consortium has recently produced a major report on London HIV paediatric care endorsing the formal development of 3 service networks (London HIV Consortium Paediatric Sub-Group - Developing Service Networks in Paediatric HIV Infection 2003). There is a lack of training programmes for medical and nursing specialists in this area. Outside London other paediatricians and adult ID specialists care for children

6. Pathways of care

There are very limited numbers of specialists in Paediatric HIV in the UK (less than 10). Due to the wide geographical areas covered and the need for families with complex medical and social needs for local care, networks of care with outreach clinics, multidisciplinary audit, and joint protocols have all developed. The multidisciplinary team has played a critical role in the maintenance of adherence and good physical and psychological health in these highly vulnerable families. Nurse specialists, psychologists, pharmacists, dieticians and physiotherapists have developed care pathways and MDT networks to support families in maintaining health.

7. Gaps and pressures

There are no major centres of paediatric HIV specialist care outside London, but there are ever increasing numbers of children with HIV living outside London. We urgently need to develop regional centres for family HIV care across the UK. CHIVA and the RCPCH are working with the commissioners to try and review this major gap in service provision.

In London the needs of adolescents and transition clinics require further development, and paediatric HIV care should be integrated into developing multidisciplinary service networks of care for children with all infectious diseases.

Metabolic Disorders

1. Definition and scope

Metabolic medicine and the treatment of patients with an inherited metabolic disease (IMD) is a small but highly specialised area of medicine. These disorders represent an important part of the growing number of genetic disorders with new diagnostic tests and new treatment possibilities. Disorders have a wide clinical spectrum including disorders that may result in sudden decompensation with coma and death e.g. fatty acid oxidation disorders and urea cycle disorders. Others may follow a slow neurodegenerative course e.g. lysosomal disorders.

Many diseases are multi-system requiring a holistic approach and assessment by a wide range of specialist medical services including neonatal and paediatric intensive care, cardiology, neurology, nutritional teams and genetic services for initial diagnosis and further management.

2. Incidence and Prevalence

The service is almost entirely a tertiary one as these disorders are generally rare, most individual disorders having an incidence of less than 1 in 10,000 births. An example is Phenylketonuria (PKU), for which there is a newborn screening programme within the United Kingdom, which is well established and successful. This condition has an incidence of approximately 1 in 12,000. Recommendations exist for the diagnostic concentrations of phenylalanine and standards of treatment and monitoring.

It is difficult to define the full scope of the service for IMD, as there is currently no nationally held data for the incidence and prevalence of different conditions. This requires the urgent development and funding of disease registers. Workload tends to be more concentrated in those areas with certain ethnic minorities because of genetic inheritance patterns.

The scope of any metabolic service is rapidly evolving with increasing recognition of inherited disease, increasing patient numbers, better survival, and an increasing expectation of health service delivery from the public. There is a constant development of new treatments for adult and paediatric populations, including the recent development of enzyme replacement therapy for lysosomal storage disorders. Many are experimental and require close monitoring.

3. Expressed Needs

a) Medical Services

These should be provided in a Regional centre and coordinated by a Consultant Paediatric

specialist in Metabolic disease.

Shared care with local services is very important as many of the patients may decompensate becoming acutely unwell and need to be admitted locally. Individual emergency treatment regimes should be developed and provided for the child so treatment can start immediately when they are admitted to the local centre. There should be access to further advice from the Regional centre 24 hours a day. The patients may need other local services and in particular services for children with neurodisability.

b) Other Clinical Services

The Regional centre will provide a multidisciplinary team to support the patient with IMD. In the treatment of IMD patients an experienced dietitian plays an important role in the multidisciplinary core team. The dietary management of IMD is complex.

In addition support from clinical nurse specialists should be available. This coverage at present is variable throughout the United Kingdom.

The considerable burden imposed on families with the diagnosis, poor cognitive outcome, long-term chronic illness and sometimes bereavement means there is a considerable need for the availability of clinical psychology services as part of a multidisciplinary service. For many of these disorders, neuropsychometric assessment is the main outcome measure and is normally obtained by clinical psychologists. This need is poorly provided for at present.

c) Laboratory Services

The clinical service for IMD is crucially dependent on the specialist metabolic biochemistry laboratory service. In all centres with a clinical service there is a complementary laboratory with access to a full range of specialist testing and links to neonatal screening. The National Newborn Screening Centre came into being in April 2002 and followed on from the Medical Research Council /Department Of Health funded PKU register. Standards will be developed for the various stages in the Screening Process.

Major advances in genetics and biochemical diagnosis have also highlighted the need for specialised laboratory services. New diagnostic technology in the form of tandem mass spectrometry (TMS) is being introduced throughout the United Kingdom and will change the practice of the specialty of metabolic medicine. This technology, using neonatal blood spots, leads to the possibility of more patients being diagnosed in the newborn period with extended screening. As a wide range of disorders may be detected, immediate specialist advice and counseling will need to be available throughout the United Kingdom. The most obvious candidate for inclusion is medium chain acyl CoA dehydrogenase deficiency

(MCAD). The value for screening other disorders e.g. urea cycle disorders and maple syrup urine disease is less clear. Introduction of screening impacts on the need for rapid diagnosis and management of IMD which requires clinical, laboratory, nursing and dietetic services.

d) Voluntary Organisations

Many support organisations exist and some are specific for individual diseases. CLIMB, NSPKU and the MPS Society are examples of organisations which can provide invaluable support for parents and families.

4. Current Service Provision

The Consultant service is currently patchy (only ten centres currently provide a service) and inequitable. At present only two units have more than two consultants (GOS, London and Manchester) with expansion planned in Birmingham and London. In all other units the consultants are single-handed. As a result many patients are still under the care of district paediatricians, only some being seen in outreach clinics.

5. Pathways of Care

Each local commissioning healthcare group will require a clear link with the Regional centre to co-ordinate care for patients in their area with these conditions. Those areas with a high incidence for ethnic reasons may require enhanced services from their Regional centre, such as the provision of specific outreach and joint clinics with local paediatric services.

6. Gaps and Pressures

Currently many pressures exist which lead to an inequitable distribution of service for these patients and need significant future investment in services.

The lack of a coordinated approach and appropriate services for patients with inherited metabolic disorders throughout the United Kingdom is of extreme concern to the professionals involved in these services, the patients and their carers. These problems lead to inequity of service and are particularly acute because ;

- Many children with metabolic disorders are under the care of district paediatricians with no specialist training.
- Many of the specialist consultants providing a service are single-handed. The development of networks between Regional centres will be required and need funding for expansion.
- Evidence based guidelines for investigation and management need to be developed on a National basis.

- Information for patients, parents and health professionals is limited.
- Specialist laboratory services have a major problem with inadequate numbers of clinical scientists and a looming crisis in succession, as many senior staff will retire in the next 5 years.
- Specialised adolescent and adult services for inborn errors are at best very limited and non-existent in some areas. Many adults are still looked after in paediatric units with others lost to follow up.
- Future development of neonatal screening programmes will impact on service provision for patients.

NEONATAL INTENSIVE AND HIGH DEPENDENCY CARE

(Note this section refers to medical care only. See surgical services section for neonatal surgery.)

1. Definition and scope

Units that care for babies can have a variety of names (neonatal unit, special care unit, neonatal intensive care unit, etc) but currently these names say little or nothing about the type of work they perform. Activity has traditionally been described as: intensive care, high dependency care, special care and normal care. The definitions of these terms have varied over time, but the most up to date definitions can be obtained from the British Association of Perinatal Medicine (BAPM) www.bapm.org. Simple, broad descriptions are given in the box below. However it is important to note that any one baby may start life normal, become ill at 12 hours (say with an infection) be needing intensive care by 24 hours and move through high dependency and special care categories in the following few days before being normal again at a week. It may be necessary to move the baby from one unit to another in order to get the appropriate level of care.

Outline definitions

Type of care	Intensive care	High dependency care	Special care	Normal care
Incidence	About 1.5 to 2.0% of all live births. However casemix variation can result in significant local distortion		About 7 to 10% of all live births	All live births
Type of intervention	a) Ventilation irrespective of the babies gestation; b) Continuous airway pressure support in the first 5 days of life; c) general supportive care to the most immature infants (e.g. those born < 29 weeks gestation) in the first 48 hours of life; d) certain specific procedures -e.g. dialysis	a) Continuous airway pressure support in babies not fulfilling intensive care criteria; b) babies < 1000g not fulfilling intensive care criteria; c) babies receiving intravenous nutrition; d) certain special situations e.g. babies with tracheostomy	A whole range of babies meet this criteria if they have needs which can not reasonably be met at home and do not fulfil the criteria for intensive or high dependency care - e.g. babies requiring intravenous therapy, or phototherapy for jaundice	Babies in this category should not be in hospital. However special circumstances may exist - e.g. in babies admitted for a special investigation who need normal care whilst an inpatient

2. Evidence for standards of care

The recent review of newborn care in England (available from www.doh.gov.uk/nsf/neonatal.htm) made it clear that :-

- a) Intensive and high dependency care should be considered specialised services;
- b) All neonatal care should be delivered through managed clinical networks based on populations of about 15 to 25,000 births (i.e. will involve multiple PCOs);
- c) Each network should designate their constituent units in terms of the type of work they should undertake. The type of unit will determine the level of staffing, equipment, general support, etc that should be immediately available. See BAPM document “Standards for hospitals providing neonatal intensive and high dependency care - 2001” (available from www.bapm.org);
- d) Each network must have sound transport arrangements in place;
- e) Staffing should be matched to the type of unit.

Similar reviews are in place or planned in Wales, Scotland and Northern Ireland.

3. Pathways of care

The network concept requires a significant number of babies to be moved to access the care they require. Therefore all units accepting referrals must have adequate facilities to support and accommodate parents for, sometimes, prolonged periods.

Intensive care units should be co-located with foetal medicine services. Babies needing intensive care frequently need access to a range of other services which should be available on site or have clearly defined routes of access. These include: neonatal surgery, specialist anaesthetic support, radiology, genetics, the full range of paediatric pathology services, dietetic services, pharmacy services, community liaison, follow up assessment and support, family and cultural support services, bereavement counsellors.

All units must be able to operate at their appropriate level 24 hours a day 365 days a year. In particular a first class resuscitation and stabilisation service must always be available in all units. There must be clear local arrangements for the follow up and continuing care of infants with ongoing problems, including transfer to appropriate hospital and community services for older children.

4. Audit, evaluation, and monitoring

A national audit scheme is being considered. In the meantime all units should collect data using the BAPM definitions and produce an annual report. Existing national mortality data produced

by ONS is difficult to evaluate (because of local confounding factors) and almost impossible at the level of the individual PCO (because of small numbers). Data from individual hospitals is similarly difficult to evaluate because of casemix differences. However these existing types of data can be supplemented by: survival rates at particular gestations (using babies alive at the onset of labour as a denominator), the proportion of babies that have to be transferred outside the local network in order to get care (should always be < 5%), and the proportion of preterm babies alive and normal at 2 years.

5. Voluntary organisations

Individual hospitals often have active parent groups. The largest national organisation is BLISS (Baby Life Support System www.bliss.org.uk).

6. Gaps and Pressures

The high pressure nature of the work and the financial rewards available have resulted in nursing staff shortages and recruitment problems for a number of ancillary groups such as specialist pharmacists and dieticians. The need to reduce doctors' hours of work is placing great strain on existing medical working patterns. It is envisaged that networks will improve this situation to some extent.

NEPHROLOGY, INCLUDING RENAL REPLACEMENT THERAPY

1. Definition and Scope

1.1 The role of the Tertiary Paediatric Nephrology Service is to improve the care offered to children with nephrological diseases, including those in whom the primary problem is Urological. The service offers expertise in the many individually rare conditions presenting to local services. It also offers diagnostic and therapeutic modalities which are not available in primary and secondary care. Diagnostic expertise is dependent on high quality paediatric renal pathology and renal imaging. There is an important preventative aspect to the service as effective management may delay the development of renal failure both in childhood and adult life. Advice about genetically determined renal disease is another preventative aspect of the service.

1.2 An important principle is to provide care as close to home as possible. Outreach clinics held jointly with the local paediatrician are a mainstay of such local care but there is evidence that the provision of these services are significantly limited by the underprovision of both medical and non-medical professionals¹.

1.3 All aspects of renal care are heavily dependent on a multiprofessional team. This is particularly important for the management of the most severe acute and chronic conditions and preventative elements of the service.

1.4 Counselling the child and family so that they have a realistic understanding of the likely progress of the disease both in childhood and adult life is a special role for the Tertiary team and again requires multiprofessional working^{1,2}.

1.5 There is a high degree of inter-relationship between paediatric nephrology, paediatric urology and transplant surgery. Other important inter-relations include:

- Paediatric Intensive Care
- Neonatal Intensive Care
- Foetal medicine
- Genetics
- Paediatric Imaging
- Paediatric Histopathology
- Adult Renal Services

2. Incidence and Prevalence

2.1 Table A lists conditions usually treated by PCO and secondary teams³. Table B shows rarer conditions usually managed wholly or largely by the regional centre.

2.2 Detailed information about the incidence and prevalence of end stage renal failure is available from the Paediatric Renal Registry⁴. These reports have identified that ethnicity has a very major impact on the incidence and prevalence of end stage renal failure. Acute Renal Failure (ARF) has an incidence of 197/million total population /year. After infancy the incidence of ARF is 8/million population/year. Nephrotic Syndrome (NS) has an incidence of approximately 1:6000 children under 16 years of age. In 90% of cases of NS there will be no risk of renal failure but even in this good prognosis as many as 60% may require involvement of the Tertiary Centre at some time. The 10% in whom there is a risk of renal failure will require specialist follow up. The incidence of nephrolithiasis is approximately 2 children/million total population/year.

2.3 Table C outlines the additional low volume services which need to be provided for large populations.

Table A – Conditions which may be handled locally or by shared care

Uncomplicated Urinary Tract Infection (1% boys, 5% girls)
 Nocturnal Enuresis (15% at 5 years)
 Daytime Enuresis (1.5% at 5 years)
 Antenatal Hydronephrosis (1:125)
 Chronic Renal Failure (Mild/moderate)
 Acute Renal Failure not requiring dialysis
 Uncomplicated Nephrotic and Nephritic Syndrome
 Haematuria
 Proteinuria
 Mild to Moderate Hypertension

Table B – Uncommon Conditions

Acute renal failure requiring dialysis
 Severe Chronic Renal Failure
 Chronic Dialysis and Transplantation
 Complicated Nephrotic Syndrome
 Severe or chronic glomerular disease
 Vasculitis (except mild Henoch-Schonlein Syndrome)
 Chronic tubulointerstitial disease including Fanconi Syndrome
 Renovascular Hypertension
 Severe hypertension and hypertensive crises
 Nephrolithiasis

Table C – Low Volume Service Provided for Large Populations

Haemodialysis – acute and chronic
 Peritoneal dialysis – acute and chronic
 Access for haemo and peritoneal dialysis
 Plasmapheresis
 Renal Transplantation – Cadaveric and Living Related
 Tissue typing and renal immunology
 Renal biopsy
 Urodynamics
 Specialised Renal Psychosocial support including education
 Feeding clinic including specialised paediatric dietetics
 Specialised renal pharmacy

3 Current Service Provision^{1,2}

3.1 There are currently 9 centres in the UK which provide Nephrology, Dialysis and Renal Transplantation for children (Belfast, Birmingham, Bristol, Glasgow, Great Ormond Street, Guy's Hospital, Leeds, Manchester, Newcastle and Nottingham).

3.2 Cardiff (linked with Bristol), Liverpool (linked with Manchester), and Southampton (with Guy's) provide Nephrology and Dialysis services with links to centres indicated for transplantation.

3.3 Leicester and Sheffield (with Nottingham), Oxford, and Edinburgh (with Glasgow) provide Nephrology services.

3.4 All tertiary centres, supported by Nephrology/Dialysis centres, operate care networks with secondary services. Outreach clinics are an essential feature of these networks and workforce underprovision seriously limits the provision of outreach services¹.

4 What children Need

4.1 The necessary concentration of low volume services in a small number of centres inevitably means travel for at least some aspects of management. This may be over long distances and for haemodialysis the child/young person will have to travel three times per week. Transport must be reliable and timely. All children under 16 years will need to be accompanied. For children travelling frequently, particularly those travelling three times a week for haemodialysis, it is often not practical for the parent/guardian to take on this role and a 'carer' will need to be provided to take on this task.

4.2 Renal failure management is very demanding and there is significant psychological morbidity and social cost for the child and family⁸. To mitigate these burdens on the child and family and to maximise the life opportunities of the patient and siblings it is essential that adequately resourced specialist renal psychosocial services are available to advise the family and engage local services.

4.3 A particular concern is the provision of adequate educational support and advice to minimise the impact of frequent admissions to hospital and, for children on haemodialysis, the impact of three times a week treatment in hospital. This requires teaching when in hospital and liaison with the local schools^{1,2}.

4.4 It has been shown that transition to adult care has particular problems for young people with renal problems, particularly those with renal transplants⁵. The transfer to adult services of young people needs careful planning.

5. Gaps and Pressures

5.1 A number of gaps and pressures were identified during preparation of the recent re-

ports^{1,2}.

5.2 There is an imminent crisis in consultant staffing. There have been a number of unfilled posts in the recent past. To avoid this in the future trainees have been recruited to a national training scheme. The first batch of trainees will complete training this year and currently the majority will not have consultant posts to apply for. They will be lost to paediatric nephrology unless an urgent solution is found to this problem. This will be a massive disincentive to further trainees applying for national training posts and the prospects for paediatric nephrology will be even worse than before the national training scheme was initiated. The modest and realistic plans set out in the Workforce Planning Document¹ will not be achievable and units will have to close. This will increase the travelling for children and their families. There will also have to be a reduction rather than an increase in outreach clinics further reducing access and increasing inequity in the service.

5.3 There are major gaps in the number of nurses. Only seven units are able to provide specialist nursing 24 hours a day and this limits the treatments available in some units¹. There are also insufficient training places for paediatric renal nurses¹.

5.4 Unfortunately the same problems arise with the staffing levels for all the other members of the multiprofessional team particularly dieticians, psychologists/psychiatrists and social workers¹.

5.5 There is a further problem with funding of paediatric renal social workers. The majority are charitably funded often on short term contracts. Furthermore those with Local Authority funding are under threat¹.

6 Organisations consulted

6.1 The organisations consulted in preparation of the recent reports are detailed in those reports^{1,2}.

6.2 The National Service Framework for Nephrology⁶ has consulted widely with patients and carers.

6.3 There has been extensive consultation with the British Renal Society, the Kidney Patient Association, the National Kidney Research Fund, the Renal Association and the British Association for Paediatric Nephrology.

7 Audit and Evaluation

7.1 All centres report their data to the Annual Paediatric Renal Registry⁴.

7.2 All transplant data is reported to UK Transplant which audits outcomes⁷.

7.3 All centres audit their activities against the Royal College Document 'Treatment of Adults and Children with Chronic Renal Failure; Standards and Audit Measures'⁸.

7.4 It is hoped that the above document plus the standards established in the recent review of units² will allow regular multiprofessional peer audit of units to be established.

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NON-MALIGNANT HAEMATOLOGY

1. Definition & Scope

Haematology is a specialist service provided in tertiary regional centres usually alongside paediatric oncology and provided by multidisciplinary teams. The aim is to provide specialist clinical and laboratory diagnosis and interpretation in rare disorders which include coagulopathies (encompassing thrombophilia), haemoglobinopathies, haemophagocytic syndromes, chronic haematological disorders (neutropenia, thrombocytopenia and haemolytic processes), unusual anaemias and provision of comprehensive care for haemophilia. Many regional centres also provide bone marrow transplant (BMT) services for non malignant indications e.g. haemoglobinopathy, severe aplastic anaemia, metabolic disorders, haemophagocytosis etc. Tertiary paediatric haematology is a low volume service for rare and difficult diagnoses provided for large populations. There is often a requirement for inpatient support for definitive treatment, blood product transfusion, iron chelation therapy and intravenous access management. Closely linked services include: immunology, oncology, blood transfusion medicine, pain services and genetics.

2. Incidence & Prevalence

Many conditions which would be appropriately managed by this service are relatively common and may be managed by primary and secondary services with co-ordination and expertise provided by tertiary haematology. Others may be exceedingly rare and require extensive tertiary diagnostic and clinical input. Disorders associated with specific geographic ancestry require more resources dependant upon distribution of ethnic minority populations. The beta thalassaemia gene carriage in the Pakistani population is between 6%-10% and the sickle haemoglobin gene carriage can be up to 25% in some Afro-Caribbean populations. Tertiary centres often provide an annual review process in conjunction with secondary centres where most care may be provided.

3. Expressed Needs

As for any rare disorders children and their carers require prompt access to diagnostic expertise and explanation of their investigations, results and diagnoses and a clear and coordinated plan for management including emergency access to care. There is a requirement where possible to provide shared care with community based or DGH services. Information regarding links to other agencies (education, social services) and support networks (e.g. Shwachman Diamond syndrome, Fanconi anaemia, ITP support groups) is available. User groups are beginning to define what they expect from healthcare providers and are developing standards of care for

management with specific medical and multidisciplinary input. The UK Sickle Cell Society and the UK Thalassaemia Society are developing such standards.

4. Evidence for Standards of Care

The British Committee of Standards in Haematology (BCSH) as part of the British Society of Haematology (BSH) has commissioned and published guidelines for standards of care in all areas of haematology which include management of ITP and blood transfusion in children (www.bcshguidelines.com) The UK Haemophilia Centre Directors Organisation (UKHCDO) have produced guidelines for the management of head injuries in haemophilia. Registries for very rare disorders (severe aplastic anaemia, Fanconi anaemia, dyskeratosis congenita, Diamond Blackfan anaemia, chronic neutropenias etc.) are managed by enthusiastic individuals and reported to the Paediatric Sub-Committee of the BSH. Additionally a directory of laboratories and contacts providing specific specialist investigation for rare disorders is available on the BSH website.

5. Current Service Provision

Multidisciplinary tertiary haematology care is usually provided alongside oncology with clinical nurse specialist involvement. Some centres provide outreach clinics at DGHs jointly with paediatricians. On-call arrangements aim for twenty-four hour availability of advice, though this will depend upon numbers of haematologists locally. On-call is often shared with oncology. In large centres a separate non malignant on-call rota may exist. Facilities for emergency access and care are provided e.g. for haemophilia, bleeding (platelet disorders), acute sickle crises (infarction, sequestration) and the management of serious infections.

6. Pathways of care

There is an increasing requirement from transition to adult services. Transition in haemophilia is reasonably well developed but there may be difficulties in other areas e.g. haemoglobinopathies, neutropenia and pre-leukaemia syndromes. Provision is rather rudimentary at present, a great deal of work is required to get this right which may involve shared adult and paediatric clinics.

7. Gaps and Pressures

Major problems with under staffing create service delivery pressures. A recent manpower survey by the Paediatric Sub-Committee of the BSH identified a major shortfall in trainees in paediatric haematology for new, vacant and retirement haematology posts. Additionally the national neonatal haemoglobinopathy screening programme which is coming on line in April 2004 will provide additional pressures for all paediatric haematologists in the UK.

ONCOLOGY AND MALIGNANT HAEMATOLOGY

1. Definitions and Scope

Service provision for children with oncological and malignant haematological conditions is provided by the United Kingdom Children's Cancer Group (UKCCSG) and the United Kingdom Childhood Leukaemia Working Party (UKCLWP). The UKCCSG is a national, multidisciplinary organisation which aims to advance the care of children with cancer through clinical research (web site <http://www.ukccsg.org/>). The ultimate objective of the group is to improve the outcome of children with poor prognosis malignancy and maintain excellent rates of cure, whilst reducing long term toxicity for those children with a good prognosis disease. The (UKCLWP) is responsible for the care of children with malignant haematological conditions and functions in parallel and complements the UKCCSG. The UKCCSG has a network of nearly 500 members from 22 United Kingdom treatment centres. Within the UKCCSG there is multidisciplinary representation (chemotherapy, pathology, radiology, radiotherapy and surgery).

2. Incidence and Prevalence

The incidence of malignancy (including leukaemia) is 1 in 600. Therefore in a PCO with an under 15 year population of 50,000 about five children would be expected to present with a malignancy each year.

Overall, in the United Kingdom each year 1,200 children present with a malignancy, 30% with leukaemia and 27% with central nervous system tumours. Studies have clearly demonstrated that the probability of long-term cure is increased when patients are treated at a UKCCSG centre (Stiller, British Journal of Cancer 1994) and treated according to an established protocol (Stiller et al, British Medical Journal 1990).

3. Evidence of Standards of Care

The treatment for childhood malignancy is by national and international protocols. Currently there are 37 protocols covering the majority of childhood malignancies.

NICE guidelines are being established for childhood malignancy and the UKCCSG and Childhood Leukaemia Working Party are actively involved in this process.

4. Current Service Provision

Eighty seven per cent of children with malignancy in the United Kingdom are treated at the 22

UKCCSG centres where multidisciplinary care is delivered. At most of the UKCCSG centres shared care is established with Paediatric Oncology Shared Care Centres (POSCUs). Guidelines are established for interaction between a UKCCSG centre and a POSCU. A detailed survey of current service provision with a number of core staff in each centre is currently being established.

At each UKCCSG centre there is an established multidisciplinary team involving a paediatric oncologist, surgeon, radiotherapist, pathologist, radiologist, specialist nurses, physiotherapy, occupational therapy, pharmacy, dietician, psychologists and psychiatrists.

There are a number of national organisations involved in supporting children with malignancy and their families, including the Childhood and Adolescent Cancer Partnership (CACP), the National Alliance of Childhood Cancer Parent Organisation (NACCPO), Sargent Cancer Care for Children, Macmillan Cancer Relief, Cancer Leukaemia in Childhood (CLIC) and the Teenage Cancer Trust.

The majority of UKCCSG centres also focus on the care of adolescents with malignancy and strongly interact with the Teenage Cancer Trust. These teenage units provide interaction with adult services to ensure a seamless delivery of care through childhood, adolescence and adulthood.

5. Pathways of Care

Clear referral pathways are established between district general hospitals and the UKCCSG centres when a diagnosis of childhood malignancy or a central nervous system tumour is suspected.

6. Gaps and Pressures

Survival for childhood malignancy overall is 70%. This has been achieved by multi-modality care and the treatment of children according to national and international protocols. The survival for some previously poor prognosis malignancies, for example in acute myeloid leukaemia, has been achieved by the use of intensive chemotherapy. Currently, the United Kingdom results for this type of leukaemia are highest in the world. As a consequence of this the demands for intensive support for children undergoing these treatments is increasing. A survey is being undertaken of the current needs at UKCCSG/CLWP centres. The key themes which are appearing are:-

- i) Limited capacity to care for children with malignancy at UKCCSG/CLWP centres due to the lack of trained medical and nursing staff. This has resulted in some children with

malignancy being transferred from one UKCCSG centre to another in order that investigations can be instigated and treatment delivered.

ii) Reliance on voluntary organisations for the provision of service. Childhood cancer charities, particularly Sargent Cancer Care for Children, Macmillan and CLIC provide funding for members of the multidisciplinary team. Due to financial variability in charitable organisations, services may be cut without warning and the vulnerability of the service has recently been demonstrated.

As 70% of childhood malignancies are treated on national and international protocols, with the introduction of Good Clinical Practice (GCP) and the EU directive for clinical trials, there are increasing regulatory requirements. This is putting enormous pressure on the UKCCSG and UKCLWP centres, in terms of compliance with trial regulations.

PAEDIATRIC INTENSIVE CARE

1. Definition and Scope

The specialty provides a service for infants and children who may benefit from more detailed observation and treatment than is normally available on standard wards and departments. This is usually taken to imply a use of technological support, including mechanical ventilation and/or invasive monitoring.

Intensive Care may be needed in patients with established or incipient organ failure, arising either from an acute illness or as a predictable phase in a programme of treatment (for instance following cardiac surgery).

The appropriate centralisation of Paediatric Intensive Care (PIC) facilities has increased the need for specialist transfer teams to move critically ill children from the healthcare facility in which they initially present to the Paediatric Intensive Care Unit (PICU).

2. Incidence and Prevalence

Approximately 2% of children admitted to hospital will require Intensive Care. There is evidence that the use of intensive care by children is increasing, and there are 2.3 admissions to intensive care per thousand children per year or more. There is marked seasonal variability in PICU bed use. Although the provision of one bed per 30,000 children would cover most of the year, at times of peak demand this increases to one bed per 16,000 children.

A number of specialist services depend upon PICU provision. The range of admission diagnoses seen in one PICU is shown in the box.

Respiratory
Neurology/neurosurgery
Generalised Sepsis
Cardiothoracic disease
Trauma and burns
Neonatal Surgery
Other Surgery
Nephrology
Hepatology
Oncology/haematology
Metabolic Disease (inc Diabetes)

The range of diagnostic groups admitted to a tertiary Paediatric Intensive Care Unit.

3. Expressed Needs

Families need rapid access to the knowledge and skills of the PICU team, either directly or by proxy through their local clinicians. This must be available at a senior level at all times and emphasises the need for strong networks between PICUs and their referring hospitals. Families need safe, timely and accurate management of their critically ill child, by clinicians who are trained and competent in intensive care, in facilities appropriate to their needs. Communication with parents and patients should be clear and honest, with agreed management plans and dis-

charge arrangements where appropriate.

4. Evidence for Standards of Care

This can be found in the Children's NSF; 'Paediatric Intensive Care "A Framework for the Future" Department of Health, 1997; "A Bridge to the Future - Future nursing workforce and education in paediatric intensive care" Department of Health, 1997; "Standards for Paediatric Intensive Care" Paediatric Intensive Care Society 1996; The Bristol Royal Infirmary Inquiry Report; Report of the Paediatric & Congenital Cardiac Services Review.

5. Current Service Provision

PIC is currently provided in 29 units in England and Wales. These vary from single specialty units (for instance looking after children with burns or following cardiac surgery) to larger multi-specialised units. Units dedicated to the care of critically ill children have a better outcome than units that practice intensive care on an occasional basis. Larger units may be more efficient, but the exact relationship between unit size and quality of care has yet to be defined.

6. Pathways of Care

Critically ill children will present to their local hospitals that should have the facilities and expertise to resuscitate and stabilise them. The PICU should liaise with its referring hospitals to provide appropriate training and advice to facilitate this. There should be clear lines of communication between referring centres and the PICU both for admissions and at the time of discharge. There should be agreed protocols for management, admission and discharge. Following discharge there should be appropriate arrangements for follow up, and if necessary for counselling of the parents in the event of the child not surviving.

7. Gaps and Pressures

Significant advances have been made following the centralisation of PIC services, but further progress is hampered by a lack of appropriately trained children's nurses. Medical staff expansion is needed to allow appropriate reduction in working hours for medical staff. Increasing difficulty is being found in appointing appropriately experienced middle grade medical staff.

These staffing pressures are exacerbated by the need for a continuous emergency and out of hours service.

Pressure on intensive care facilities will come from the increasing survival of technology depen-

dent children, who often are cared for inappropriately in intensive care as there are inadequate resources to allow them to be nursed at home or on a paediatric ward.

Transition

There is some overlap in care at both ends of the age spectrum, with some PICUs caring for term or near term infants who in other areas would be cared for by neonatology services. Similarly young adults with, for instance, congenital cardiac disease, may be best looked after in PIC where there is the appropriate expertise and experience. In both cases, provision should be made for age appropriate facilities.

Audit and indicators of excellence

All units should participate in the PIC Audit Network (PICANET) and those undertaking cardiac surgery should also audit their work through the Central Cardiac Audit Database. Structures should also exist to allow for regular liaison with referring centres, other specialties that access PIC facilities, and parent groups.

PAEDIATRIC NEUROSCIENCES

Paediatric Neurodisability

1. Definition and Scope

The aim of tertiary neurodisability services is to provide specialist expertise to children and their families in partnership with secondary and primary health services, education, social services and the voluntary sector. The system of managed clinical networks and care pathways for different disabilities recognises that some aspects of disability need specialist services to be funded as a care package. Specialist neurodisability services occur at secondary and tertiary level and it is through locally managed care pathways that the relative contributions may be negotiated in order that the child has access to the following specialist components of care which may be diagnostic, management strategies or actual services. Specialist neurodisability services include:

- Rehabilitation following acquired brain injury
- Surgery for cerebral palsy, scoliosis, gait analysis
- Spasticity management including botulinum toxin
- Severe cognitive and behavioural regression in epilepsy
- Complex communication disorders, diagnosis and therapeutic intervention
- Mixed complex learning problems often with neuropsychiatric co-morbid symptoms
- Provision of communication aids
- Sensory impairments e.g. cochlear implants
- Services for severe visual and hearing impairment
- Specialised seating/wheelchairs and orthoses

The precise package will vary from district to district depending on a number of factors but budgetary provision needs to be made to purchase specialist services from the tertiary care provider. Children and their families need locally organised multidisciplinary care through childhood to adult life. Needs are likely to change over time with key moments at transition to school and adult services. A care plan is needed, shared with child and family. Involvement with specialist services may be of variable frequency throughout childhood. Collaboration across services is vital in promoting a service around the child and family. Good communication between tertiary or specialist services, other agencies and secondary level services is costly in time and needs resourcing as part of a purchased package.

Tertiary neurodisability works closely with paediatric neurology and neuropsychiatry and there is considerable overlap in type of problem and provision.

2. Incidence and Prevalence

In a typical PCO with a child population under 16 years of 50,000, there will be at least 1,000 (2%) of children with **moderate/severe** disabilities (including 0.3-0.5 % with severe learning difficulties) and there will be 10-20% or more with **milder** problems but still affecting learning or behaviour.

Autistic spectrum disorder	3-6/ 1000	150-300/ PCO
Cerebral palsy	3/ 1000 live births	120/ PCO
Mod. learning difficulties	3/ 100	1200 / PCO
Epilepsy	0.5 / 100	250 / PCO
Neuromuscular Disease	0.5 / 1000	25/ PCO
Severe learning difficulties	0.3 / 100	150 / PCO
Acquired brain injury	18 /100,000	9 / PCO
Prevalence of each disease/disease group / PCO of 50,000 children <16 years		

Box A

Many children will have dual disabilities, rare disabilities and/or comorbidities and have needs for specialist services. Some of these are outlined in Table C in the Neurology section of this document. Two other examples of care provision are given here.

Pathways of care provision: two examples

	Cerebral Palsy	The Providers			
		Voluntary Agency	Primary	Secondary	Tertiary
The pathway	Identification		Parent, HV, GP, Neonatal team Paed. physio.	Diagnosis Child development services	Neurodisability and/or Paed.neurologist
	Assessment	SCOPE, Contact a family	Social support	Multidisciplinary team	Specialist MDT / surgical /orthotic/neurodisability services
	Management	SCOPE	Counselling Inclusion in primary care health services including preventive health/dental measures	Therapy, play, Treatment e.g. of GOR / epilepsy Paed. surgery Liaison with education, housing, leisure	Feeding/communication aids Gait analysis/seating Physio/botulinum toxin Paed orthopaedic Surgical services for limbs, joints and spines Liaison with Special Education Seating

(Also see over)

	Communication disorder	The providers			
		Voluntary agency	Primary	Secondary	Tertiary
The pathway	Identification		Parent, HV, GP, Speech/lang therapist	Child development services Mental health services Neurodisability Speech/lang therapist.	Neurodisability services for difficult diagnosis/second opinion Paed.Neurologist for co-morbidities/investigation Neuropsychiatry service for complex behaviour
	Assessment	Contact a Family, Afasic National Autistic Society	Social support	Multidisciplinary team	Specialist MDT
	Management	National Autistic Society Sibling/family services	Counselling Responsive awareness to needs of family members Promotion of liaison	Therapy, play, 'Early Bird' Specialist speech & language therapy Multi-agency coordination and liaison Continued responsiveness to changing needs	Liaison through sharing specialist teaching/speech & lang.therapy/other interventions/medication /psychology treatments etc. Continued responsiveness to changing needs

3. What children and families need

Standards

Education Act 1998 Code of Practice 2000

Standards for Child Development Services – A guide for commissioners and providers BAACH / CDDG 1999

Recommendations for minimal Standards of health Care in children with cerebral palsy Hemi--Help 1999

Disability Discrimination Act 2000

Community paediatric workforce requirements to meet the need in the 21st Century. BACCH Working Group 1999

Quality standards for children with visual impairment DfES 2002

Developing intervention/support for deaf children and young people DfES 2002

Together from the Start: practical guidance for professionals working with disabled children and their families DfES 2002

National recommendations for management of Autistic Spectrum Disorder 2003 and National Plan for autism (www.nas.org.uk)

The business plan: service development for children with neurodisability, Standing Committee on Disability RCPCH 2003 (www.cddg.org.uk)

NICE Guidelines on management of ADHD 2001

Getting the right start: National Service Framework for children – Standard for hospital services and Emerging Findings DoH 2003

NICE guidelines for management of epilepsy – to be published 2004

National Service Framework for Children-full report due 2004

Box B

All children and families want a prompt response to concerns, a coordinated response to therapy and care needs, and have an expectation of competence in health service staff. Guidelines for services and standards are in Box B.

4. Staffing - workforce requirements for service delivery

Tertiary paediatric neurodisability services should be provided by a multidisciplinary team working with paediatric neurology and neuropsychiatry. At secondary level, all districts should have a paediatrician specialising in neurodisability but the level of specialty that the multi-disciplinary team can offer will vary from district to district but ideally should contain all components (Box C).

Multidisciplinary Team (MDT)	
Core	Supporting
Consultant Paediatrician in neurodisability	Paediatric ophthalmic/ ENT /audiology
Paed. physiotherapy	Paediatric neurology, Neuropsychiatry,
Clinical psychology	Mental health services for learning disability
Occupational therapy	Palliative services
Speech & language therapy	Educational psychology
Dietician	Full range of paediatric services including investigations, radiology, metabolic etc
Paediatric orthopaedics for cerebral palsy	Genetics
Scoliosis service/seating /wheelchair services	Neurosurgery
	Box C

The particular team for each child varies according to the disability.

Other agencies fundamentally involved in provision of good neurodisability services include Education and Social Services, parent and patient user groups and voluntary agencies.

5. Gaps and Pressures

- Equity of access to tertiary services
- Need to acknowledge the cost of supporting specialist services including eg. specialist neuroradiology opinion.
- Lack of paediatric therapists and psychologists
- Lack of behaviour support
- Mental health services for learning disabled

- Very limited academic posts in the specialty
- Lack of adult services to whom transition of care can be made
- Lack of funding for equipment
- Equity of pay across therapy services

There is a shortage of skilled clinicians in the paramedical therapies eg speech therapy and a severe lack of supportive mental health services for learning disabled children. Neurodisability is a new specialty and like paediatric neurology experiences a shortage of trained doctors. Changes in educational provision, notably inclusion has placed significant pressures on therapy delivery in schools. Lack of respite services places major pressures on families. The need to develop and deliver training and services including transition of care to adult services are significant resource issues for specialist service packages.

Research priorities

- Prevention and treatment of mental health problems in learning disability
- Evidence based therapy interventions
- Developing and auditing care plans for disability
- Effective interagency working
- Cerebral palsy treatments e.g. does postural management prevent scoliosis?

Box D

Neurology

1. Definition & Scope

The aim of the Tertiary Regional Centre based Paediatric Neurology service is to improve the quality of care offered to children with neurological disorders through accurate diagnosis and cost-efficient and effective management. The service offers expertise in the many individually rare conditions frequently presenting to local services and diagnostic equipment not available in primary or secondary

<p>Headache</p> <p>Epilepsies and related disorders</p> <p>Learning disability, including specific learning disability and autism</p> <p>Cerebral palsy</p>
<p>Table A – range of conditions for initial assessment and treatment by the PCO secondary tier paediatric team.</p>

care. There are essential close links with departments of Inherited Metabolic Disease, Genetics, Specialised Children’s Pathology and Neuroradiology to aid diagnosis and with Paediatric Neurosurgery and Child and Adolescent Mental health services. A principle is to keep the care of children as close to their homes as possible. Outreach clinics held jointly with local paediatricians have hitherto supported informal clinical networks to help fulfil this aim. The British Paediatric Neurology Association is currently working with the Health Service Modernisation Agency to establish a formalised network for the management of epilepsy as a first step in establishing managed networks for other aspects of paediatric neurology. Details are available at www.bpna.org.uk There is a close liaison with District hospital and community-based paediatricians.

2. Incidence and prevalence

Table A shows conditions usually treated by PCO primary and secondary tier teams. Paediatric Neurologists also treat these disorders at their regional centre base to set standards of care, maintain expertise and facilitate training of General Paediatricians and Neurodisability consultants. Table B illustrates the range of conditions likely to be seen by the Paediatric Neurology service per annum. Table C summarises the additional services that these children may require.

3. What children need

Irrespective of the condition or age of the child, parents and children tell us that they want timely and accurate diagnosis and assessment by a credible person (including recognition of associated impairments that are not immediately obvious); clear honest understandable information; a negotiated and agreed intervention plan; guidance on the role of and links to other agencies including education, social services and parent organisations; support for child and family through the school years; a transition plan for adult life.

Seizure disorders not responding to first –line medication
 Children with epilepsies requiring Neurosurgery
 Movement and sleep disorders
 Neuromuscular Disorders
 Brain tumours
 Non-traumatic coma, including meningitis
 Stroke
 Degenerative disease/metabolic disorders
 Multisystem disease involving other tertiary specialists
 Neuro-intensive care involving 1 in 10 ICU beds
 Mental illness presenting with physical symptoms

Table B – Uncommon conditions treated by paediatric neurologists through clinical networks. (Numbers per PCO per annum)

- Acute and medium term medical rehabilitation (acute brain injury and spinal cord injury including chronic ventilation and severe epilepsy).
- Behavioural/cognitive/neuropsychological rehabilitation programmes.
- Management of spasticity: multidisciplinary programmes including gait analysis, botulinum toxin and intrathecal baclofen in conjunction with neuro-orthopaedics and orthotic management.
- Dysphagia management and investigations including videofluoroscopy.
- Regional neuromuscular service providing diagnosis and support, aided by skilled family care officers.
- Specialist spinal/postural management.
- The diagnosis and advice on management of uncommon syndromes and developmental problems require the resources of both paediatric neurology and specialised neurodisability services.

Table C – low volume services provided for large populations; the overlap with Neurodisability services will be noted. Paediatric neurologists provide most tertiary Neurodisability services

Voluntary organisations consulted include Epilepsy Action, The Muscular Dystrophy Association, CLIMB, SCOPE, AFASIC, MENCAP, NAS, Downs Syndrome Association, and Contact a Family.

4. Staffing

Each PCO team should be part of a network and subscribe to agreed protocols and audit processes. Each should have a Paediatrician with Special Responsibility for the management of children with neurodisabilities including epilepsy. The PCO team should provide an initial assessment of all conditions in Box A. The paediatric neurology service should be involved where one of the criteria in Table D is met. The benefits of a network approach are that existing skills can be utilised and made available more widely without each PCO having to invest in expensive resources and scarce expert staff to meet the highly specialised needs of a small number of children, avoiding “distance-decay”.

Availability of staff There is a serious shortfall in service delivery due to a lack of paediatric neurologists and specialist nurses. Currently one in five PCOs lacks an outreach service. Further details are available in the BPNA Manpower document and the Epilepsy Clinical Network document published on the BPNA web site. There are plans to increase the current number of 60 UK paediatric neurologists significantly over the next 10 years.

Emergency and out of hours requirements The paediatric neurology service offers a 24

Diagnostic uncertainty	Parental/doctor request	
Failure of first-line medication	Surgery selection (e.g. epilepsy or cerebral palsy)	
Neurointensive care	Complex (more than one) disability e.g. Epilepsy with learning and behavioural difficulties	
A child with a condition in Table B or requiring a service in Table C		
<table border="1"> <tr> <td>Table D: Criteria for involving the regional neurology service.</td> </tr> </table>		Table D: Criteria for involving the regional neurology service.
Table D: Criteria for involving the regional neurology service.		

hour a day telephone service for all PCOs in its clinical network. Consultant neurologists are available for immediate recall to their regional centre to support in particular Neurointensive care except where staffing levels have to date made this impossible.

Travel Children may need to visit their regional neurology centre – usually based at the nearest large city-based teaching hospital - to undergo specialised tests or be assessed by a specialised team. The links with PCO teams should facilitate subsequent local follow up.

Transition Children with continuing neurological problems need follow up in adult life. Some specialists in adult neurology, rehabilitation or learning disability medicine will provide advice and support but few offer the coordinating role undertaken by paediatric services. This may fall to the GP if there is no other service available, and this can work well if the GP has been well briefed by the paediatric team. Transitional care clinics are now in place in many regions for young people with seizure disorders or other neurological conditions.

Audit, evaluation and indicators of excellence See the section on Neurodisability.

Further Reading

www.bpna.org for details of Manpower planning and proposals for Clinical Network develop-

ment.

A Guide for Purchasers of Tertiary Services for Children with Neurological Problems.

BPNA 1998 – available from your local department of paediatric neurology

Neurosurgery

1. Definition and scope

This specialty includes the full range of congenital and acquired conditions affecting the child's nervous system, which are amenable to surgical treatment. Congenital abnormalities include neural tube defects such as spina bifida and encephaloceles, hydrocephalus and a variety of other conditions that affect the skull and face (craniofacial disorders) and spine. Acquired conditions include head injuries, brain and spinal cord tumours, infections, vascular problems, epilepsy surgery, spinal and functional neurosurgery which increasingly may help the treatment of spasticity and cerebral palsy.

2. Incidence and prevalence

There are approximately 4,500 neurosurgical cases operated on per annum for children under the age of 16 years in England and Wales. The majority of operations are related to shunts and drainage procedures in the treatment of hydrocephalus and its complications. Surgery for head injuries varies between 4 and 10%, and tumours account for about 15% of cases.

3. Expressed needs

Several published documents indicate the needs for the neurosurgical care of children, notably produced by the Society of British Neurological Surgeons (see www.sbns.org.uk). They include "Safe Neurosurgery" (1993), "Guidance for Services for Children and Young People with Brain and Spinal Tumours" (SBNS with UKCCSG 1997), "Safe Paediatric Neurosurgery" (1998) and "Safe Neurosurgery 2000" (2000). Reference to the neurosurgical care of children is also included in "Children's Surgery - A First Class Service", prepared by the Paediatric (now called Children's Surgical) Forum of the Royal College of Surgeons of England (2000).

The objectives in these documents were to ensure that children's care would be of the highest quality, delivered by recognised Paediatric Neurosurgeons supported by the appropriate staff and facilities. This would enable children to obtain the same level of care and expertise currently available for the practice of adult neurosurgery.

Paediatric neurosurgical practice requires a close working relationship with other paediatric specialists, who include Paediatricians, Neurologists, Anaesthetists, Oncologists, Endocrinologists, Ophthalmologists, ENT Surgeons, Plastic and Maxillofacial Surgeons.

4. Evidence for standards of care

A major document is “Standards for Patients Requiring Neurosurgical Care” prepared by a Clinical Standards Committee of the Society of British Neurological Surgeons in collaboration with the Regional Specialised Services Commissioning Group (July 2002). There are also NICE guidelines related to head injuries, that include specific recommendations concerning children. The documents referred above also relate to standards. National audits exist for shunts in the treatment of hydrocephalus, management of paediatric tumours including the central nervous system (UKCCSG) and craniofacial surgery (NSCAG annual supraregional audit).

Voluntary organisations include the Association for Research into Hydrocephalus and Spina Bifida, Headlines (Craniofacial patients support group) and there are other organisations related to paediatric neurology such as MENCAP, and so on.

5. Current service provision

There are 17 neurosurgery departments in England and Wales undertaking neurosurgery in the paediatric age group. The number of cases vary from 70 to 800 cases per year. There are 36 Consultants in the UK with a major or sole interest in Paediatric Neurosurgery. Of these, only 4 (i.e. 2%) of the total consultant neurosurgical population base are pure Paediatric Neurosurgeons. In 13 Units neurosurgery is undertaken in adult and paediatric combined sites. In only 4, at Alder Hey, Liverpool, Birmingham, Great Ormond Street and Manchester is there a separate paediatric hospital where neurosurgical practice is undertaken. In these centres there are more than 400 cases operated per annum, whilst there are more than 200 cases per year operated on in 13 Units.

The essential requirements of a Paediatric Neurosurgery Service is that it should be capable of a comprehensive 24 hour service, including Consultant and middle grade surgical cover. Specific facilities include paediatric beds, paediatric intensive care, appropriate anaesthesia, paediatric nurses and onsite CT scanning.

All Neurosurgeons receive training in “core competence” particularly for emergency surgical procedures, including shunt surgery and head injuries. Specialist paediatric neurosurgical practice, with relatively small numbers of cases involved, generally are referred to a less number of centres where there is special expertise and sufficient numbers to maintain competence and allow appropriate audit. Examples include brain tumours, epilepsy surgery and surgery for complex craniofacial anomalies.

6. Pathways of care

Paediatric Neurosurgery is a tertiary service with most referrals, notably acute emergencies, coming from General Paediatricians, General and Orthopaedic Surgeons. There are protocols in most centres for the transfer of head injured children, and there is available literature on this subject. Essentially there are emergency and elective referrals. The document “The Child’s Journey with a Head Injury” by Mr. Ian Pople, Consultant Neurosurgeon at Frenchay Hospital, Bristol illustrates suitable pathways for optimising the care of children, especially with the threat from the results of a major head injury (see the SBNS web site).

7. Gaps and pressures

A major pressure at present relates to both Consultants and Specialist Registrars and their hours of duty. With the European Working Time Directive, a major issue for the short and medium term future relates to timetable arrangements both for Specialist Registrars and Consultant Neurosurgeons. This becomes a big pressure in a smaller neurosurgical department or stand alone Children’s Hospital. A second key issue is the provision of Paediatric Intensive Care Unit beds and the maintained skills and confidence of Anaesthetists when some emergency surgery is required in the local facility.

PATHOLOGY

1. Definition and scope

Paediatric pathology is concerned with age dependent disorders of children from one month of age to fifteen years of age and includes:

1. Biopsy diagnosis of surgically removed tissue from children
2. Tumour diagnosis; Treatment of childhood tumours is crucially dependent on accurate diagnosis and staging as well as ancillary investigations of removed tumour tissue
3. Autopsy investigation of children dying from any paediatric disease, particularly with congenital heart disease or cancer and of children dying suddenly and unexpectedly.

Perinatal pathology is concerned with disorders of fetal life and the neonatal period. It is primarily an autopsy-based service and provides information essential to the assessment of risks to, and the management of future pregnancies.

2. Incidence and prevalence

A typical PCT with an annual live birth rate of 3,000 would expect to have roughly 15 stillbirths and 10 neonatal deaths. At current autopsy rates approximately half of this number would be expected to have a post-mortem. Numbers of children with major surgical problems or with tumours are likely to be much smaller e.g. in the UK every year there are approximately 100 children under 15 years of age who develop Neuroblastoma - one of the commoner childhood malignancies.

3. Expressed needs

These are well laid out in the Joint Working Party Report on the Future of Paediatric pathology.

4. Evidence of Standards of Care:

The Working Group Report of 2002 recommended that pathology services for children should be provided only by paediatric pathologists concentrated at specialist centres. The joint report of the Royal Colleges on Fetal and Infant Pathology recommends perinatal pathology should be carried out only by those with adequate training and facilities. Paediatric and perinatal pathology provides an audit of obstetric and neonatal care. Recent concerns have also been raised regarding the diagnosis of the causation of death in infancy.

The Royal College of Pathologists has recently published Guidelines on Autopsy Practice and is

shortly due to publish minimum data sets for paediatric and perinatal autopsy.

4. Current Service provision

In 2002 there were 26 Pathology departments in the UK that provided specialist paediatric pathology.

5. Pathways of Care

Paediatric and perinatal pathology lies across many pathways of care. Pathology is vital to the investigation and treatment of childhood disorders including childhood cancer. Perinatal pathology is the only validated method of definitively establishing causes of perinatal and infant death and thus an audit of obstetric and neonatal care.

6. Gaps and Pressures

There is currently a shortage of consultants in paediatric pathology with many vacancies left unfilled. Entry into the specialty at SpR level is disappointingly low and shortages are likely to continue for the foreseeable future.

References

The Future of Paediatric Pathology Services: Report of the Joint Working Group, RCPCH 2002

CESDI 8th Annual Report, 2000

UKCCSG Scientific report 2002

Guidelines on Autopsy Practice. Royal College of Pathologists, September 2002

Joint Working Party Report on Fetal and Perinatal Pathology - Royal College of Obstetricians and Gynaecologists and Royal College of Pathologists, June 2001

RESPIRATORY MEDICINE

1. Definition and scope

Respiratory disorders in children are common and diverse. The wide range of problems is listed in the National Definitions Set¹. Paediatric Respiratory Medicine has important links to many other specialties, including intensive care, thoracic surgery, ENT, immunology and allergy, gastroenterology, neonatology, neurology, oncology, orthopaedics, radiology and anaesthetics. Other essential disciplines include nursing, physiotherapy, dietetics and psychology.

2. Incidence and prevalence

Respiratory conditions are the commonest cause of paediatric hospital admission, accounting for 14% of UK hospital admissions² and over 50% of long term illnesses in children.³ Asthma is the commonest cause of school absence, CF the commonest lethal inherited condition. Table A shows the incidence / prevalence of respiratory conditions for a standard PCO with 50,000 0-14 year olds and a birth rate of 3,000. Many common respiratory illnesses can be well managed in

Table A. Prevalence of respiratory conditions in a "standard" PCO

Primary chronic conditions

	Children affected	Notes
Asthma	10,000	150 admissions per annum Often managed by non-specialist
Cystic Fibrosis	20	
Chronic lung disease of newborn	3 p.a. going home on oxygen	
Non-CF Bronchiectasis	10-15 (<i>est</i>) (of whom 3 will have Primary Ciliary Dyskinesia)	
Sleep disordered breathing	50 children with significant problems	
Long term ventilation	1-2	
Rare serious lung disease	1-2 (<i>est</i>)	
Congenital lung and airway disease	3 p.a.	

Complications of other conditions

Neuromuscular	12 with severe disease	
Neurological	50 (<i>est</i>) with severe cerebral palsy or degenerative conditions	
Immune deficiency	2 (<i>est</i>)	
Oncology	Uncertain	
Chest wall and scoliosis	Uncertain	

Acute admissions

LRTI	160 p.a.	Often managed by non-specialist
Croup		Often managed by non-specialist
Empyema	2 p.a.	

Notes. *est*=Estimated. Other figures are based on single centre audits or published prevalence data

Table B. Low volume services provided for large populations

- Bronchoscopy
- Home oxygen and monitoring
- Diagnostic pulmonary function testing
- Management of respiratory complications in neuromuscular disease
- Management of recurrent aspiration problems
- Investigation and treatment of recurrent lower respiratory tract infections
- Diagnosis and treatment of sleep-disordered breathing
- Home ventilation

primary and secondary care. However, those children with severe common illnesses such as asthma or pneumonia, or with one of the many rarer respiratory conditions require the involvement of a specialist multidisciplinary team. “It is important to ensure that the diversity of respiratory conditions is recognised and the skill base to manage and diagnose this diversity is maintained.”⁴

Factors likely to affect prevalence

The incidence of allergic disease and asthma has increased over the last 20 years and this rise may continue. Changing expectations from parents and professionals are increasing the demand for more intensive respiratory intervention in neurological diseases. It is now recognised that several conditions, such as primary ciliary dyskinesia, non-CF bronchiectasis and sleep disordered breathing are currently underdiagnosed, leading to preventable morbidity.

3. Expressed needs

Several patient organisations have stressed the need for consistent standards of care. The National Asthma Campaign recently published a Patient’s Charter⁵ and the CF Trust has set standards of care⁸. An alliance of professional and patient groups produced “Bridging the Gap”, a document setting standards for commissioning services for respiratory conditions⁴. The PCD and TOF Associations have endorsed guidelines⁹. Patients with conditions such as sleep disordered breathing or chronic lung disease of the newborn are disadvantaged as they do not have such well organised patient groups.

4. Evidence for standards of care

Standards of care for asthma are defined in the SIGN/BTS guidelines⁶, and NICE guidance on inhalers⁷; the Cystic Fibrosis Trust has published evidence-based standards of care⁸; European standards exist for PCD⁹ and for respiratory care in neuromuscular disorders; there are interna-

tional standards^{10,11} published for the management of sleep disordered breathing in childhood and UK standards are being developed by a RCPCH working party; there are guidelines for long term ventilation¹².

5. Current service provision

Workforce information is updated annually by the British Thoracic Society¹³. There are marked inequities in levels of staffing. For example, in 2001 the number of paediatric respiratory consultants varied from 5 in Liverpool to 1 in Cardiff, and the number of specialist nurses from 5 in Leicester to 1 in Manchester. For some diseases, such as CF, these inequities are being increasingly recognised. However, services for other patient groups, such as children who require home ventilation or the assessment of sleep related breathing disorders shows marked (and indefensible) inequalities of access. It is difficult to commission and fund these low volume, high cost services, which are often established on an ad hoc basis, without planning or funding. Inadequate service provision leads to unnecessarily prolonged hospital stays, the “blocking” of intensive care beds, and unnecessary stress on families.

Respiratory services are very dependent on many other specialties (see above). High quality care requires a multidisciplinary team with adequate numbers of respiratory function technicians, physiotherapists, specialist nurses, dieticians, social workers and psychologists.

6. Pathways of care

There are nationally approved care pathways for asthma⁵ and for Cystic Fibrosis⁶. Pathways are being developed by the RCPCH working party on Sleep Physiology. Asthma has been used as an exemplar of patient empowerment with the introduction of written self-management plans, agreed with the patient or carer.

7. Gaps and Pressures

In many areas, paediatric respiratory medicine remains understaffed and under resourced. Inequity of access remains a major problem due to workforce inequalities and a lack of agreed funding processes for more unusual lung diseases (see above). Few, if any, centres, can provide an out-of-hours service within European Working Time Directives. The current national shortages of trained paediatric radiologists is of great concern as high quality imaging is an essential aspect of assessment and diagnosis. Many regional centres have inadequate lung function testing facilities. In the past the CF Trust has provided considerable financial support for CF teams but due to changing priorities within the Trust, this funding is likely to be withdrawn in the near future.

Foreseeable crisis development

There are two areas of particular concern:

sleep-disordered breathing and home ventilation services - prevalence increasing rapidly; increased awareness of diagnosis and intervention, by both parents and professionals. New technology has made intervention more feasible. The increasing prevalence of obesity is leading to increased obstructive sleep apnea. No agreed method of funding; current services often based on local enthusiasm and inadequate ad hoc funding.

Respiratory care of children with neurological disorders - changing parental and professional expectations has led to an increase in the intensity of respiratory support provided. These children are living longer, occupying more intensive care beds, and becoming more technology-dependent. Respiratory paediatricians should help lead the provision of care for these patients, but there are no plans/funding to take account of this.

Transitional care arrangements

The transition into the respiratory service for babies with neonatal conditions such as chronic lung disease of the newborn is inconsistent, with some home oxygen services provided by the neonatologists and some by respiratory paediatricians. Readmissions are common and it is imperative that a seamless service is provided for these patients. The transition between paediatric and adult respiratory services is also very variable. CF services generally have transitional arrangements, but other conditions such as asthma, bronchiectasis, congenital problems, and sleep-disordered breathing frequently require continuing specialised care in adult life and this is often less well provided.

References

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RHEUMATOLOGY

1. Definition and Scope

Children and young people with musculo-skeletal or multi-system rheumatic diseases are seen within the Paediatric Rheumatology Service. The common conditions are sometimes managed locally, whilst the rarer conditions may only be seen by the tertiary specialist team. It is important to note that many of the children will have challenging multi-system disease and that when they present with acute illness their management may be very complex, involving other sub-specialties, including intensive care.

2. Incidence and Prevalence

Table A shows that the inflammatory rheumatic disorders are relatively rare in childhood. Children should be referred to an experienced paediatric rheumatology service for initial assessment. In larger trusts, this may be provided locally but in smaller ones there may be a need for

<p>Management of common musculo-skeletal and soft tissue syndromes in the growing child (anterior knee pain, hyoer mobility)</p> <p>Management of acute, sub-acute, medium and long term joint disease, including joint replacement (e.g. From <1yr to 18+)</p> <p>Management of childhood vasculitis and connective tissue disease (cytotoxic care and stem cell transplantation)</p> <p>Management of severe acute multi-system disease, often requiring intensive care support</p> <p>Multi-agency management of chronic pain and fatigue syndromes within a care pathway, integrating primary, secondary and tertiary care</p> <p>Diagnosis and management of uncommon syndromes and unusual symptom complexes (e.g. skeletal dysplasias)</p> <p>Assessment, prevention and management of osteoporosis in children (i.e. secondary to steroid, not only for rheumatology patients)</p> <p>Assessment and management of children with joint aches and pains</p>

Range of Conditions (Table A)	Approximate number per PCT of 50,000 children
Juvenile Idiopathic Arthritis, sub groups inc. oligo and polyarthritis	50-75
Juvenile Idiopathic Arthritis, systemic	1-2
Connective Tissue diseases (dermatomyositis, scleroderma, SLE)	5-10
Systemic Vasculitis and acute multisystem illness	5-10
Severe inflammatory eye disease	2-5
Chronic Fatigue Syndrome and ME	50- 100
Severe localised and generalized pain syndromes	25 -50
Genetic / Dysplasia and other conditions	20 - 50
Mechanical / Orthopaedic	400 - 1000
Osteoporosis prevention and treatment	30-80

tertiary referral centre . A few patients will need to attend a national center. A Clinical Network can facilitate management of some of these conditions, so that children with less challenging management need not then attend the tertiary centre. There is a significant workload associated with non-inflammatory disorders in childhood. While some of these can be managed by a PCO based service once the diagnosis has been made, others may require very specialised multi-disciplinary management, including Child and Adolescent Mental Health.

3. Expressed Needs

Irrespective of the condition or age of the child, parents require:

- Timely and accurate diagnosis
- Clear, honest information about the condition and the outlook
- A negotiated and agreed strategy for management
- Guidance on the role of and links to other agencies, including education, social services and parent organisations
- Support for the child and family through the school years and into adolescence with a transition plan into adult services for adult life.

4. Evidence for Standards of Care

There is a statutory duty for a designated liaison doctor to provide educational advice in respect of assessments for special educational needs and to liaise with schools over the children's problems. Standards for care of children with disabilities can be found in the NSF and on the following website: www.rcpch.ac.uk.

Voluntary organisations that may be involved with individual families include CCAA, Arthritis Care, Lady Hoare Trust and Contact a Family.

Facilities required (Table B)

Specialist musculo-skeletal diagnostic facilities – clinical, radiological (i.e. Ultrasound, MRI, CT and dexta scans) and laboratory
Access to day case ward for intravenous treatments (i.e. Cyclophosphamide, Pamidronate and biologic therapies)
Access to Theatre for procedures, i.e. joint injections and biopsies (including use of Entonox, sedation and anaesthesia)
Facility for multi-disciplinary and multi-agency assessment and management of patients
Specialist Physiotherapy and Occupational Therapy resources (i.e. hydrotherapy and splint making)

5. Pathways of Care

As much treatment as possible for this group of children should be provided at a local centre. A complete multi-disciplinary / multi-agency team is necessary at the tertiary centers, which will need to support those working locally in secondary and primary care across the managed clinical networks. There should be agreed protocols and audit processes across each managed clinical network, and there should be access to the specialised services as listed in Table C. In

Specialised Services required (Table C)

Shared care with other specialty services especially Ophthalmology, Orthopaedic Surgery, Nephrology, Cardiology, Dermatology, Child and Adolescent Mental Health, Maxillofacial, and Neurology as well as transition care to adult Rheumatology.
Specialist Allied Health Professional team, including Nursing, Physiotherapy, Occupational Therapy, Podiatry, Orthotics and Dietetics for assessment, patient education, treatment and rehabilitation programmes.
Specialist liaison with Education and Social Services to address needs of specific chronic illness.
Specialist emotional and spiritual support to meet the needs of the family and child with chronic illness.
Expertise in assessment of neglect, emotional abuse and child protection for children with chronic illness and disability.
Staff with expertise in the role of complementary therapies for the specialist conditions treated .

some parts of the country these specialised services may be combined on one site, but in others they may be dispersed. The benefits of a network approach are that existing skills can be utilized and made available more widely without each PCO having to invest in expensive resources and scarce expert staff to meet the highly specialised needs of a small number of children.

6. Gaps and Pressures

Availability of Staff / Access to Clinical Networks

The outcome of patients with inflammatory rheumatic problems has been radically altered by the use of intra-articular steroids, immunosuppression and biologic therapies. However, these developments - which are mainly day case, outpatient and community-based - have not been funded uniformly to allow all patients access to robust clinical networks. There is a major shortfall in staffing required for compliance on all aspects of NICE guidance 35 and other guidelines for immunomodulatory treatment. Current resources are insufficient to maintain drug costs, medical, nursing and audit standards.

Emergency and Out-of-Hours Requirements

There increasingly needs to be 24-hour access to expertise in managing the acutely ill child, and

those immunosuppressed and on biologic therapies. Rheumatic emergencies can be very complex and demanding, often requiring intensive care facilities. Current regional services need to explore ways of achieving this within even wider networks, especially in less densely populated areas. Most Tertiary Paediatric Rheumatology Services are currently provided by single handed specialists without funding for a second consultant. These are geographically appropriate services but this shortage compromises routine specialist care for 52 weeks each year as well as emergency cover.

Travel

Children may need to visit a distant Centre in order to undergo specialised tests or be assessed by a specialised team, or to gain access to a wide range of specialties. Links with local units and outreach clinics should facilitate follow-up, so that the child does not need to travel to a distant centre every time they require review. Many of these children have pain and mobility difficulties, so unnecessary travel should be avoided. School absences should be minimized.

Transition

Most children with rheumatic disease need follow-up in adult life. Some specialists (including nurses) with a major interest in adolescents, will provide advice and support during this period. The organisation of the transfer to Adult Services will be managed from Paediatrics and requires enthusiasm, training and commitment from adult colleagues. The primary health care team will need to be involved. Whilst some regions have well developed transition services in other areas, there is no such service as recommended by RCP / RCPCH working party report and requested by young people.

7. Audit, Evaluation and Indicators of Excellence

- Registers of diseases and their presentations
- Protocols and pathways for management of newly diagnosed children with Rheumatic conditions
- Evidence of the ability to monitor the effects of newer therapies
- Evidence of regular liaison meetings within the service, and if necessary with other agencies
- Involvement of a user group (parent organization, formal or informal)
- Evidence of Network Development linking to providers of specialised services, such as the Ophthalmic and Orthopaedic aspects of Rheumatic disorders
- Evidence of systems in place to support families and children
- Evidence of participation in research

Specialised Child and Adolescent Mental Health Services

1. Definition & Scope

The specialty includes:-

1. mental health aspects of conditions primarily managed by paediatricians such as chronic or life threatening physical illness (e.g. diabetes, cystic fibrosis, chronic inflammatory bowel disease, cancer, neurodegenerative disorder, sequelae of mild or severe head injury)
2. mental health disorders requiring active management by paediatricians because of the consequences for physical health (anorexia nervosa, self harm)
3. conditions where aetiology & management generally cross boundaries and shared management is common (e.g.. somatisation, chronic fatigue syndrome, encopresis)
4. neurodevelopmental disorders which may also be categorised as neuropsychiatric disorders (autistic spectrum disorder (ASD) including Asperger syndrome, Attention Deficit Hyperactivity Disorder (ADHD), Tourette syndrome, specific and global learning disability)
5. neuropsychiatric disorders often coexisting with neurodevelopmental disorder (eg mood disorders, conduct and oppositional disorders (CD/ODD), obsessive compulsive disorder (OCD), self injurious and other challenging behaviours, disorders of emotional control, sleep disorders, selective mutism, post-traumatic stress disorder (PTSD), schizophrenia and psychotic disorders.)

Table A: Conditions within the scope of a district or PCO-based team

	Prevalence /1000	No cases per PCT (0-14yr pop. 50 000)
Chronic /Life threatening conditions	0.2	10
Anorexia Nervosa	1-2	50-100
Self Harm	0.2	10
Neurodevelopmental disorders		
ASD	6-9	300-450
Tourette/chronic tics	1	50
OCD	5-10	250-500
ADHD	10-30	500-1500
Specific LD	100	5000
MLD/SLD ie. IQ<70	25	1250
Neuropsychiatric disorders		
Emotional disorders (including depression, anxiety, OCD, phobia. PTSD)	43	2150
CD/ODD	53	2650
Psychotic disorder	0.3	15 (schizophrenia only. No data bipolar disorder)

Sources: ONS, Mental Health Of Children & Adolescents In Great Britain 2001, Practical Child Psychiatry, Lask, Taylor, Nunn, BMJ publishing, 2003,

Table B Uncommon conditions

Pervasive refusal
Severe psychiatric disturbance in early /pre-adolescence requiring in patient facilities
Psychiatric conditions in hearing impaired (approx 25-30% of deaf children)

Table C low volume services provided for large populations

Eating disorders
Psychiatry of hearing impairment
CFS/ME
Complex psychopharmacological management of ADHD, ASD, TS etc
Forensic psychiatry in Learning disability, ASD/high functioning autism
Mental health (in particular, challenging behaviours and emotional disorder) in learning disability, ASD/high functioning autism
Management of infants in Perinatal Psychiatry units

2. Expressed Needs

From parent organisations e.g. Attention Deficit Disorder Information Service, National Autistic Society

Families' views on services for autism, Dept of Public Health, Fife NHS Board, 2001

3. Evidence for Standards of Care

NICE -except for ADHD, Conduct Disorder and Depression, the guidance below refers to management of adults but makes some reference to children

- *Attention Deficit Hyperactivity Disorder (ADHD) Methylphenidate*, 2000 (Due to be reviewed 2004 as guidance for all drugs used in ADHD)
- *Conduct disorder in children. Parent-training/education programmes* due 2004
- *Anxiety* due 2004
- *Bipolar disorder* due 2006
- *Depression in children* due ?2005
- *Disturbed (violent)behaviour* due 2004
- *Eating Disorders* due 2004
- *OCD* due ?2005
- *PTSD* due ?2005
- *Self harm* due 2004

SIGN GUIDELINES

- *Attention deficit and hyperkinetic disorders in children and young people* 2001

RCPCH

- *ADHD*

ROYAL COLLEGE OF PSYCHIATRISTS COLLEGE RESEARCH UNIT

- *Mental Health needs of children looked after*, 2000

NATIONAL AUTISM PLAN FOR CHILDREN, *National Initiative for Autism: screening and assessment*. NAS, 2003

4. Current service provision & Pathways of care

Services are provided at different levels as set out in the National Service Framework. There are important interdependencies with other specialties, requiring effective liaison and linkages.

Care Pathway and service needs for children and young people with autistic spectrum disorder, Baird et al., 327 (7413):488 Data supplement, 2003 bmj.com

Integrated pathways of care for pre-school and school age children with autism Fife Hospitals NHS Trust (unpublished)

5. Gaps and Pressures

There has been an enormous increase in diagnosis of Autistic spectrum disorder which suggests current prevalence rates are *very* much in excess of most published figures. Some evidence suggests additionally higher rates related to ethnicity and/or migration, which may have implications for services in different locations. ADHD as a treatable component of complex behavioural disorder is under diagnosed. Access to services in both ASD & ADHD often restricted, low numbers of practitioners, long waiting lists up to 18-24 months. Delay in diagnosis is unacceptable as there are recognised benefits of early intervention. Conduct disorder represents a large proportion of referrals to CAMHS although evidence based effective treatment can be delivered by non clinical professionals.

- Mental Health diagnostic facilities for some groups e.g. young offenders is restricted.
- Mental health problems in Asperger syndrome/high functioning autism inadequately recognised and often poorly managed by generalist services. Related forensic services under resourced.
- Lack of services for children and young people with disabilities, e.g. with self injurious behaviour.
- Lack of suitable inpatient facilities and provision for management of acute crises.
- Inadequate levels of specialists in neuropsychiatry and neuropsychology and unfilled psychiatry posts.

- Longstanding unresolved issues relating to transition to adolescent/adult services with no consistent agreed age of transfer between services (varies between 14 and 19yrs.) Frequent lack of expertise in adult services in conditions with developmental aetiology.
- Great potential for delivering many services through non medical staff. Relevant issues around these area are in the NSF emerging findings (new style mental health workers) and Green Paper (restructuring the workforce)

Paediatric Anaesthesia

1. Definition & Scope ¹

Paediatric anaesthesia encompasses: provision of anaesthesia for neonates, infants and children covering a wide range of surgical conditions and sub-specialities; the control of acute and chronic pain; provision of advice on safe sedation for children undergoing investigative procedures; and involvement in paediatric resuscitation services. All units where children are treated must be able to resuscitate and stabilise collapsed / critically ill infants and children. This applies to hospitals with Accident and Emergency Departments providing unrestricted access to children. Anaesthesia in children should only be undertaken where there are support services, equipment and facilities appropriate to the anaesthetic needs of children; also, specially trained anaesthetic and nursing staff whose paediatric skills are employed sufficiently frequently to maintain competency with children.

Table A – range of children’s anaesthetic service provision at non-specialist hospitals	
<ul style="list-style-type: none"> • Anaesthesia for a range of simple elective or emergency surgical procedures / investigations in otherwise healthy children of all ages, who are not requiring specialist surgery. • Sedation and anaesthesia for investigative procedures e.g. MRI, CT, BSER. • Acute pain service. • Resuscitation of the collapsed / critically ill child, stabilisation and institution of intensive care; this includes children who present following major trauma. • High Dependency Care 	
TableB Circumstances requiring more specialised anaesthetic expertise	Table C Low volume services for large populations
<p>All neonates requiring surgery / anaesthesia. Anaesthesia for infants with significant co-morbidity. eg: the ex-preterm neonate; infants with history of severe neonatal IRDS, chronic lung disease, pulmonary hypertension, and long-term oxygen therapy/respiratory support (Neonate= Age 0-4/52 : Infant = Age 4/52-12/12) Anaesthesia for other children with significant co-morbidity. Eg: metabolic disease; significant congenital heart disease; major respiratory disease; complex syndromes particularly those involving the airway, breathing or circulation (excluding Downs); serious neuromuscular disorders, unstable epilepsy; major fluid and electrolyte disturbance.</p> <p>Anaesthesia for:</p> <ul style="list-style-type: none"> • major orthopaedic surgery (especially if significant blood loss is likely eg, corrective spinal surgery) • neurosurgical procedures • cardiac investigations, interventions or cardiac surgery • complex ENT surgery (especially if surgery or detailed assessment of the upper airway (ie.larynx/ trachea) is required) • major plastic and burns surgery • kidney, liver or bowel transplantation • Any child who requires post-operative intensive care 	<ul style="list-style-type: none"> • Neonatal Intensive Care • Paediatric Intensive Care • Paediatric Critical Care Retrieval Service • Chronic Paediatric Pain Service • Paediatric ECMO <p>Children requiring specialised surgery or procedures as set out in the Specialised Services National Definitions Set (2nd edition)-Specialised Services for Children-Definition no 23 (SectionD, pp23-29) under the headings:</p> <ul style="list-style-type: none"> • Specialist paediatric surgery • Paediatric orthopaedic surgery • Paediatric ophthalmology • Paediatric cardiac/thoracic surgery • Paediatric ear nose and throat surgery • Paediatric oral/maxillofacial surgery • Paediatric plastic and burns surgery • Paediatric urology • Complex child/adolescent gynaecology • Paediatric oncology • Paediatric neurosurgery

Incidence and prevalence

Children may undergo anaesthesia in *District General Hospitals, Tertiary Paediatric Units/ Children's Hospitals, or in Single Speciality Tertiary Units.*

Table A outlines the service, which would normally be delivered by a district anaesthetic team.

Table B indicates conditions and situations requiring more specialised expertise: this is normally only available in tertiary units and in a few larger district hospitals.

Table C specifies the tertiary services and linkages required in centres providing this expertise.

What children need

Child and family-centred care is required: parents should be involved; they should normally be able to be present at induction of anaesthesia and during recovery. In all clinical areas, children should be managed separately from adults, in an environment that is child friendly and has appropriate equipment, facilities and staffing.

2. Planning

District Hospitals providing anaesthetic, surgical or emergency services need to produce clear guidelines that define the extent and nature of children's surgery and anaesthesia they can provide. This should take account of anaesthetic departmental structure, staffing, expertise and available competencies; the availability of elective and emergency surgical expertise for children; on-site surgical sub-specialities; paediatric medical support; age and other thresholds for transfer to a larger, more specialised or tertiary unit. Transfer thresholds may be lower in smaller district hospitals where the provision of children's anaesthetic and surgical services may be more limited. Standards of anaesthetic and peri-operative care should comply with Royal College guidelines on the provision of paediatric anaesthesia services, irrespective of where care is delivered.¹

The numbers of children who require anaesthesia/surgery justify each PCO establishing a team to ensure that their needs are met. Each team should incorporate a range of paediatric expertise (anaesthetic, surgical, nursing etc.), be part of a regional network; and establish the availability, nature and quality of children's anaesthetic services both locally and regionally. They will also need to establish the location of specialised services listed in Table C. In many parts of the country these are centralised onto a single site to allow concentration of scarce resources and staff. A network approach should ensure that existing well-established services are utilised for those local children requiring more specialised management in addition to promoting close links between tertiary and non-tertiary centres. Additional funding will be needed if a child requires paediatric intensive care following complex surgery.

Availability of staff

Children should be anaesthetised or have their care supervised by consultants or other career grade anaesthetists with appropriate training and competencies in paediatric anaesthesia. Anaesthetic trainees must be supervised and have unimpeded access to a consultant. At all times, anaesthetists will need assistance from staff (nurses, operating department practitioners) with paediatric skills and training. Following anaesthesia, children should be managed in a post-anaesthesia care unit/area by staff who undergo regular paediatric resuscitation training. Post-operatively, members of staff, trained and competent in paediatric resuscitation, should always be immediately available.

Emergency and out-of-hours requirements

In tertiary paediatric units/children's hospitals, a consultant paediatric anaesthetist together with a resident middle grade trainee will provide on-call cover. In non-tertiary units, out-of-hours provision will depend on capability/size of each unit, together with availability of a suitable surgeon, anaesthetist and back-up staff/services. Some larger non-tertiary units may have sufficient numbers of anaesthetists with paediatric skills/competencies to provide a separate consultant rota for children; in most others, it will be the consultant on-call for the rest of the hospital who will provide paediatric cover. Some of these consultants may have no regular daytime exposure to paediatric cases although they will, in the past have received some instruction in paediatric anaesthesia. It is particularly important to ensure that these individuals are kept up-to-date and competent in paediatric resuscitation, in case they are required to advise or participate in the resuscitation of a collapsed/critically ill child. Specific funding should be made available to subsidise these aspects of CME/CPD.

If out-of-hours emergency surgery is required in a child, care can be delivered locally if the operation is relatively minor, the anaesthetic straightforward and the requisite skills, competencies and facilities are available. If the anaesthetist has limited paediatric competencies, the case should be judged on its merits. For example, it should be unnecessary to transfer healthy older children to undergo simple procedures such as wrist manipulations, suturing of lacerations etc. In addition, a significant amount of so-called "emergency" surgery in children can wait until the next day when it is likely that there will be paediatrically competent anaesthetists available; it also avoids unnecessary evening and night time operating.

Travel

Children may need to visit a distant centre in order to undergo specialist surgery electively or as an emergency. When acutely or critically ill children need urgent transfer to a tertiary centre for paediatric intensive care, they will normally be transferred by a retrieval team from the PICU; arrangements will also be required to ensure that the parents are transported, eg with the child or in a taxi.

Audit, evaluation and indicators of excellence

Units providing anaesthetic services for children should undertake audit which relates to paediatric anaesthesia. Examples of paediatric anaesthesia audit projects and quality indicators are listed in the document “Raising the Standard”.²

References

1. Guidance on the provision of paediatric anaesthesia services. London; Royal College of Anaesthetists, 2001.
2. Raising the Standard: a compendium of audit recipes for continuous quality improvement in anaesthesia. London: Royal College of Anaesthetists, 2000.(www.rcoa.ac.uk)
3. Your child’s general anaesthetic. In: Raising the Standard: Information for Patients pp 105-108. London: Royal College of Anaesthetists; 2003 (www.rcoa.ac.uk)

Children's Surgical Services

Paediatric surgery is a regionally based specialty. In the UK and Ireland there are 27 units delivering a paediatric surgical service. Many are sited in regional teaching/ training centres. On average each serves a population of about 2 million but there is a wide range from 500,000 to 5 million.

1. Definition and scope

The specialty of *Paediatric Surgery* includes not only *general paediatric surgery* but also *subspecialty paediatric surgery*, namely

- Neonatal surgery
- Paediatric gastrointestinal surgery
- Paediatric urology
- Paediatric surgical oncology
- Paediatric thoracic surgery
- Paediatric neurosurgery

The developing specialty area of antenatal /fetal diagnosis and surgery for congenital structural anomalies is demanding an increasing commitment of the time and expertise of the neonatal surgeon.

A recent document "Reconfiguration in Paediatric Surgery" (British Association of Paediatric Surgeons, Feb 2003, <http://www.baps.org.uk/bapspublications.htm>) outlines proposals for development of the specialty. It forms part of the Senate of Surgery of Great Britain and Ireland's review of the need to reconfigure surgical services nationally.

2. Consultant staffing

It must be emphasised that paediatric surgery is a consultant-based specialty. Trainees at SHO and Specialist Registrar (NTN, VTN or FTN) level are supernumerary and do not contribute in a significant way to service delivery. A regional unit will normally consist of general paediatric surgeons each with subspecialty interests and commitments and specialist paediatric urologist(s).

Consultant numbers are currently about 130 but expansion is proposed. This is essential to allow increased consultant cover to compensate for the effects of European Working Time Directive and Junior Doctors Training requirements and to provide 24 hour consultant-based emergency cover.

3. Standards Documents

A variety of documents consider the future, remit and standards of paediatric and neonatal surgery:

- *The Provision of General Surgical Services for Children*. Senate Paper 3. The Senate of Surgery of Great Britain and Ireland. July 1998.
- *Surgical Services for the Newborn*. British Association of Paediatric Surgeons/ The Royal College of Surgeons of England. June 1999.
- *Children's Surgery - a First Class Service*. Report of the Paediatric Forum of the Royal College of Surgeons of England. May 2000.
- *Paediatric Surgery Standards of Care*. British Association of Paediatric Surgeons. Nov 2002.

British Association of Paediatric Surgeons (www.baps.org.uk)

All Great Britain and Ireland paediatric surgeons are members of this professional association. Working in close association with the four Surgical Royal Colleges, it plays a leading role in matters relating to professional affairs, teaching and training, setting standards, ethics, audit and research. Formal subcommittees are established in the fields of: Trauma; Research and Clinical Outcomes; Education and Training; Ethics.

The Association website links to both UK/ Ireland and international /Overseas organisations and provides a service for dissemination of information and debate and discussion for the 900 world-wide members.

Ophthalmology

1. Definition and Scope

The specialty includes a range of very common disorders, which can and should be managed on a local basis. There is scope for much of strabismus and amblyopia assessment and management to be carried out by community teams of Orthoptists and Optometrists working to robust guidelines and with fast track access to hospital services when problems arise.

Much rarer sight threatening conditions, several of which overlap with neurological disorders, require an expert team approach centred on a sub-specialty service

2. Prevalence

TABLE A

Incidence/Prevalence

Strabismus	2% population
Amblyopia	2% - 4% population
Epiphora	20% population (with 5% requiring treatment)
Cataract	3-4 per 10,000 births
Glaucoma	1 per 12,000
Retinal dystrophy	Rare but responsible for 10% of visually handicapped children
Visual problems of prematurity	Up to 50% of <750gm would need treatment (Stage 3)
Non-Accidental Injury	Varies with area: up to 50-60 per 1,000,000 pop'n per year
Neuro-Ophthalmic disorders	Individually rare but commonest cause of visual handicap
Non-Organic Visual Loss	?
Uveitis	?

What children need

Families need rapid and convenient access to skilled professionals in order to deal with the more common problems. Models of care provision within a community setting are available in the DoH document "Meeting the challenge: A strategy for the Allied Health Professions, November 2000". For the less common problems they require an early assessment and then care within dedicated sub-specialty units, where supporting services are available. These services include neurology and neurosurgery, oncology, neurophysiology, genetics, anaesthesia, rheumatology, as well as the necessary equipment infrastructure.

Voluntary Organisations

RNIB, Retinoblastoma Society, SENSE, RP Society

3. Current Service Provision

Services for the common disorders are generally local and good, but tend to be centred on hospital premises. Dispersing them to community facilities should be examined on a wider scale. High quality sub-specialty services are available in all regions.

4. Pathways of care

Children gain access to the services through:

Family doctor service, Paediatricians, Ophthalmologists not specialising in Paediatric Ophthalmology, from established community services and from other disparate groups such as the visiting teacher service, support organisations etc.

5. Gaps and Pressures

- There is a national shortage of Orthoptists which is exaggerated in the South East of England because of the cost of living in that area.
- The value of visual screening continues to be debated. If community screening were to be abandoned then referral from other sources less well informed than Orthoptists would probably lead to an increased Out Patient load. It might also lead to later referral with the implication of slower and less complete response to treatment.
- In some regions children requiring frequent hospital attendance e.g. those with cataract, are involved in time consuming and expensive travel.
- Access to imaging and neurophysiological testing can be difficult, leading to delays in establishing a diagnosis and instituting appropriate treatment and support.

Oral and Maxillofacial Surgery

Oral and maxillofacial surgery (OMFS), oral surgery, oral and facial surgery and facio-maxillary surgery are the terms used to express the type of surgery that is undertaken by a Department of Oral and Maxillofacial Surgery.

Oral and maxillofacial surgery is a surgical discipline, which deals with the diagnosis, evaluation and treatment of conditions arising on the face, in the jaws and mouth and their environment, including the maxilla, mandible, nose, neck and salivary glands in all ages, paediatric and adult. Treatment includes the operative, non-operative, intensive care management and rehabilitation for this complex area.

OMFS is unique in that it requires training in both dentistry and medicine. The aim of the specialty is to provide a comprehensive diagnostic and surgical service, often in a multidisciplinary manner, to our medical and dental colleagues

Specialised maxillofacial surgery:

1) Orthodontic dentoalveolar surgery

Common problem for every PCO

2) Paediatric trauma (facial lacerations -**very common**, facial bone trauma - **fairly common**, dental trauma - **very common**) Treatment is provided in Regional / Sub-regional units.

3) Cleft lip and palate provided by cleft surgeons in Regional Cleft Centres in a multidisciplinary team (CSAG Recommendations) 80 - 100 cases / year / centre

4) Paediatric Craniofacial Surgery - under discussion at present

Paediatric craniosynostosis correction is provided in collaboration with Paediatric Neurosurgeons usually in Regional Neurosurgical Units. **15 - 30 cases / year / centre**

Syndromic Paediatric Craniofacial Surgery is provided in the NSCAG Designated Supra-Regional Units These conditions are **uncommon**.

5) Paediatric maxillofacial oncology is provided in a multidisciplinary Head and Neck team, often working in collaboration with a paediatric surgeon. These conditions are **uncommon**.

6) Temporomandibular ankylosis: This is a specialised maxillofacial area and is usually a sub-specialty interest in a maxillofacial Unit 2- 5 cases / year / 3,000,000

7) Drooling (salivary): This is a specialised and under-resourced area and is usually a sub-specialty interest in a Maxillofacial Unit. Close collaboration with rehabilitation and neuro-disability units is required. **Common problem** treated by medicine, positional devices and surgery if conservative treatment fails.

2. Evidence for Standards of Care

The recommendation from the Royal College of Surgeons is for a Consultant to population ratio of 1:150,000. It is essential that Consultants should not work in single-handed units and therefore Units should cover as a minimum a population of 300,000. Referral is directly to the Regional / Sub-Regional Unit. The face is of paramount importance to the child and family and irrespective of the condition or age of the child, parents need a timely and accurate diagnosis, easily understandable and clear information, an agreed treatment plan and access to support groups for the particular condition - facial deformity.

- 1) Clinical Effectiveness Committee BAOMS www.baoms.org.uk
- 2) Clinical Effectiveness Committee Faculty of Dental Surgery
www.rcseng.ac.uk/dental/fds/clinicaleffectiveness
- 3) CSAG Report Cleft Lip and Palate

3. Current service provision

Maxillofacial surgery is by definition a specialised service provided in University, Specialist, District General and Dental Hospitals and often provided in multidisciplinary teams.

4. Gaps and pressures

There is a genuine shortage of Consultants available to provide paediatric maxillofacial surgery.

The specialty provides support to a defined anatomical area and often the child will be supported from birth into adulthood.

Orthopaedics

Paediatric orthopaedics is, by definition, provision of orthopaedic services for children under the age of 16 in the United Kingdom. Paediatric orthopaedics falls into two main categories: elective, and the management of trauma.¹ Approximately 1 in 4 children each year will attend an A&E Department with an injury and a significant proportion of these will require orthopaedic referral and management.

Paediatric orthopaedic services are split into two main areas at present, surgeons who work in specialist (tertiary) centres and those who work in the district general hospital setting. There should be an orthopaedic surgeon in each district general hospital who takes responsibility for the management of children's orthopaedic problems. However all the orthopaedic surgeons working in district general hospitals are responsible for, and are involved in, the management of paediatric trauma.

1. Conditions which should be managed at a district general hospital:

1. Management of paediatric trauma in children under the age of 16, including fractures
2. Management of common paediatric conditions (i.e. surveillance of Developmental Dysplasia of the hip (DDH))
3. Early management of club feet
4. Assessment of normal variants
5. Management of bone and joint infections
6. Surveillance and management of children with neuromuscular disabilities (e.g. cerebral palsy)

District General Hospital Setting

There should be an orthopaedic surgeon responsible for the care of children within the district general hospital setting. His responsibilities are to look after paediatric orthopaedic conditions mentioned above. His colleagues will form a rota to manage the more common children's fractures. It is of note that 50% of children's fractures are to the forearm and can be safely managed in the district general hospital. Other more complex fractures, including those associated with neurovascular problems or associated with multiple injuries, should be referred to a tertiary centre.

Surgeons in specialist centres should not be expected to manage all paediatric orthopaedic fractures. This facility must be available in district general hospitals. Children and their parents should not be expected to travel a significant distance for the management of these relatively simple injuries. However their management must take place in a safe environment for the child.

Surgeons in district general hospitals require in-patient paediatric support, a separate ward for children, an appropriately trained anaesthetist to administer a safe anaesthetic for the fractures to be treated, appropriately trained nurses on the ward and an appropriately trained physiotherapist to aid rehabilitation. The district general hospital should have all diagnostic facilities available in the form of haematological services, clinical chemistry and radiology. The radiology department should have ultrasound, an MRI scanner, a CT scanner and bone scanning facilities.

A network should be available so that consultants in district general hospitals can liaise with the local tertiary centre and mechanisms for emergency transfer of the more complex fractures should be in place. Any child who has multiple injuries should be referred to a specialist centre.

Conditions which should be managed in specialist centres

1. Operative intervention for DDH.
2. Complex surgery in children with cerebral palsy - management requires liaison with a paediatrician with an interest in neurodisability.
3. Limb lengthening.
4. Scoliosis.
5. Bone tumours.
6. Bone dysplasias.

Specialist Centres

In the document *The Specialist Services National Definitions*² there is a section on orthopaedics. These are the conditions that should be referred to specialist centres. Many of these conditions rarely occur and, for this reason, they should be treated in tertiary centres to ensure appropriate expertise is gained. The tertiary centres have full paediatric support and anaesthetists trained to anaesthetise children from birth to 16 years. The hospital should have facilities for treating children of these ages and special specific facilities should be available for the adolescent child.

In a tertiary centre it is expected that there should be appropriate intensive care services on site. All appropriate imaging must be available on site in the form of MRI scanner, CT scanner, ultrasound and isotope bone scan. Back up in the form of physiotherapy, occupational therapy and orthotics must be available on site.

2. Pathways of Care

Managed Clinical Networks

Tertiary centres should develop links with other district general hospitals to ensure an appro-

priate referral pattern. Those children who can be safely managed in the district general hospital should be managed in that facility and only those requiring specialist services should be transferred expeditiously.

Tertiary centres must have facilities for parents to stay on site. Parents may want to stay with their child waiting for an operation and in their rehabilitation phase. If they travel a significant distance the only way this can be managed is by having accommodation within or adjacent to the hospital.

A tertiary centre should be staffed by a number of consultant orthopaedic surgeons to ensure appropriate expertise is available at all times. If a number of surgeons are appointed, areas of specific expertise can be developed within the unit.

3. Gaps and Pressures

At present there is a shortage of specialists in paediatric orthopaedics, the specialty is relatively unpopular and there are significant difficulties in filling the posts in district general hospitals. This situation is likely to continue for the foreseeable future. One solution may be to appoint an increased number of specialists in the tertiary centres and arrange a “hub and spoke” relationship with the district general hospitals. With this arrangement it may be possible to carry out out-patients and some operations in the district general hospitals. However an increase in the number of surgeons at the tertiary centres would have a significant effect on the requirements for beds and other facilities.

In conclusion paediatric orthopaedics at the present time is at a crossroads. There are significant problems with staffing in district general hospitals with people appropriately trained in the management of paediatric orthopaedics. The tertiary centres are increasingly being referred patients from district general hospitals which could be treated locally if the appropriate expertise within the orthopaedic and anaesthetic departments were present. Their workload is increasing exponentially and waiting time for out-patients and surgery is steadily increasing.

References

1. Children’s Surgery: A First Class Service. Royal College of Surgeons of London, 2000. Pages 72-76.
2. Specialised Services: National Definition Set. Department of Health. 2001.

Otolaryngology

- Routine ENT surgery in children with substantial developmental, immunological or other systemic disease
- Neonates, infants and young children unsuitable for routine surgery in a secondary care setting
- Ear surgery for complex congenital abnormalities
- Bone anchored hearing aids and prosthetic ears
- Cochlear implants
- Management of laryngo-tracheal stenosis and other airway problems
- Repair of choanal atresia
- Management of severe congenital and developmental malformations of the head and neck
- ENT aspects of head and neck tumour surgery

Table A – Range of conditions requiring specialist management

Definition & Scope The sub-specialty includes the full range of ENT conditions in children but at tertiary level it encompasses those listed above in Table A. The way in which tertiary paediat-

- Obstructive sleep apnoea in otherwise normal children under the age of 2 years
- Upper airway obstruction in children with craniofacial anomalies
- Some cases of congenital conductive hearing loss
- Microtia and meatal atresia
- Profound sensorineural hearing loss
- Choanal atresia
- Nasal cysts and tumours
- Laryngotracheal stenosis
- Laryngo/tracheo/bronchomalacia
- Congenital vocal cord palsy
- Bronchial foreign bodies
- Subglottic haemangioma
- Recurrent respiratory papillomatosis
- Laryngotracheoesophageal cleft
- Lymphangioma of the head & neck
- Head & neck tumours

Table B - Uncommon conditions

- Paediatric anaesthesia
- Neonatal and paediatric intensive care
- Paediatric respiratory medicine
- Paediatric general surgery
- Paediatric cardiology and cardiothoracic surgery
- Paediatric imaging and interventional radiology
- Paediatric audiology
- Paediatric oncology
- Paediatric pathology
- Paediatric speech & language, voice and swallowing therapy
- Teachers of the Deaf

Table C - Support services

ric otolaryngology services are organised and provided varies considerably in different parts of the UK, and some elements may be available in local hospitals.

Incidence and prevalence Table B illustrates the rare conditions which may call for more specialised expertise and Table C summarises the support services that these children may require.

Expressed needs Irrespective of the condition or age of the child, parents tell us that they want timely and accurate diagnosis and assessment; clear honest information; a negotiated and agreed intervention plan; a short waiting time for surgical treatment; guidance on the role of and links to other agencies including speech and language therapy, education, social services and parent organisations; support for child and family through the school years; and a transition plan for adult life in those cases where treatment has not been completed by adolescence.

Voluntary organisations to be consulted include the National Deaf Children's Society (NDCS - www.ndcs.org.uk), Aid for Children with Tracheostomies (ACT - www.ACTfortrachykids.com) and various other self-help and patient support groups formed by parents for children with specific syndromes and disorders (www.patient.co.uk/selfhelp.asp).

Pathways of care Each PCO team should be part of a network and subscribe to agreed protocols and audit processes. Each should have access through ENT surgeons in secondary centres to specialised tertiary paediatric otolaryngology services backed up by the support services listed in Box C. In most parts of the country these specialised services will be combined on one site, but in some they may be dispersed. The benefits of a network approach are that existing skills can be utilised and made available more widely without each PCO having to invest in expensive resources and scarce expert staff to meet the highly specialised needs of a small number of children.

Gaps and pressures Shortages of trained Paediatric Otolaryngologists, Registered Children's Nurses (especially in Theatre and ICU), Speech and Language Therapists etc...

Emergency and out of hours requirements Full 24/7 cover essential.

Travel Children may need to travel some distance to reach the nearest tertiary centre. Links with local secondary teams should facilitate follow-up so that children do not need to travel to a distant centre every time

Transition Children with some ENT problems need follow up in adult life. Examples include children with otological sequelae of cleft palate, bone-anchored hearing aids, cochlear implants,

and tracheostomies for continuing airway obstruction. They will need referral to an appropriate adult centre.

Audit, evaluation and indicators of excellence Maintenance of research and audit databases, evidence of network development linking to providers of secondary ENT care, liaison with other agencies and with parent support groups.

Further information is available from the British Association of Otolaryngologists - Head & Neck Surgeons (www.entuk.org)

Plastic Surgery

There are currently about 225 consultant plastic surgeons working in the NHS. Almost all are involved in the management of children. General plastic surgical practice undertaken by the majority of plastic surgeons includes the management of trauma and the treatment of common congenital and vascular anomalies.

Within the specialty it is usual for surgeons to subspecialise in the treatment of particular conditions across the paediatric and adult age-range. Very few plastic surgeons specialise specifically in paediatric plastic surgery. However, children feature prominently in some areas of sub-specialisation. For example, surgeons with a major interest in cleft lip and palate or genital surgery operate on a larger number of children.¹

1. Definition and scope

The casemix treated by Plastic Surgeons is varied and the volume of interface work with other specialties is unknown. The conditions treated include

Specialist

- Cleft Lip and Palate (As part of a multidisciplinary team)
- Congenital Head and Neck deformities (E.g. Haemangioma, dermoids, Hemifacial Microsomia)
- Craniofacial (In Tertiary Centres - NSCAG service)
- Burns and post Burn Reconstruction (Centres, Units and Facilities)²
- Paediatric Microsurgery for complex reconstructions (as part of a team)
- Hypospadias (overlaps with Paediatric Urology)
- Epispadias, Intersex
- Head and Neck Oncology (as part of a team)

Generalist

Some of the following conditions may be simple - others may be more complex and require specialist care.

- Congenital Ear (accessory auricles, prominent ears, microtia)
- Facial Trauma
- Oculo-Plastic (Overlaps with Ophthalmology- e.g. Ptosis)
- Congenital Hand Surgery (Extra digits to complex hand anomalies)
- Hand Trauma (overlaps with hand surgeons)
- Congenital Skin Lesions

- Vascular Anomalies

9% of all NHS Children's Surgery is carried out by Plastic surgeons and 60% of this is emergency work. The majority of the elective surgery is performed on a day case basis. The more complex, requiring in patient stay, is often dealt with by subspecialists (such as congenital hand surgery) or part of a team (such as Head and Neck Oncology and Cleft Lip and Palate).

2. Incidence and prevalence

The range of conditions treated has a wide range of prevalence. Some Craniofacial conditions are as rare as 1:20,000 births, Clefts of the lip and palate are seen in 1:700 live births, Hypospadias 1:500. Haemangiomas are the commonest childhood tumour and mostly are treated expectantly. Accessory auricles and digits are common conditions treated very simply on a day case basis. Emergency surgery includes commonly finger tip injuries, tendon and nerve injuries, facial trauma and burns.

3. Expressed needs

Paediatric Plastic Surgery has links with many other surgical and non-surgical disciplines. For example, cleft lip and palate surgeons have close links with orthodontists, speech and language therapists, oral and maxillofacial surgeons, and others. Those involved in genital reconstruction have close links with paediatric nephrologists and urologists. Surgeons involved in congenital hand surgery work closely with orthopaedic surgeons and hand therapists. Those involved in the care of paediatric burns work closely with specialists in many paediatric specialties, including intensive care. Problems exist where these services are not on the same site. It is hoped that service developments following the NBCS will address these issues.

4. Current service provision.

Within Plastic Surgery Units (PSUs) the paediatric caseload may be dealt with by all consultants, subspecialists (such as hand, burns, cleft) or internal referral may take place for some or all of the paediatric cases. Compliance with regulations about children's environments and anaesthesia has tended to segregate this work. The SAC in Plastic surgery specifically identifies paediatric plastic surgery as a need for any training rotation. Plastic surgery for many simple conditions is kept at DGH level where appropriate resources allow thereby attempting to avoid the log jam of inward referral to the Hub in a network for minor cases. However the moves in Paediatric Anaesthesia drive this. Most of the surgery is very simple (equivalent perhaps to grommet insertion in ENT or dental extraction or exposure in Maxillo Facial). PSUs may be located in DGHs, Teaching Hospitals or more isolated units historically at the Hub geographically of the population they serve but not necessarily centred where all support services are located.

A good example of how well tertiary services are being managed in one region has been the response to the reconfiguration of Cleft Lip and Palate services in the Eastern part of the country. Following the Clinical Standards Advisory Group recommendations that the number of centres should be radically reduced from 57 to approximately 10-15 centres - there has been a smooth transition of care with the evolution of a network. Five teams have been amalgamated into one with all the surgery taking place on a single site with all paediatric support services including PICU. Outreach clinics are held in every previous locality so for children and their carers the only difference has been the need to travel for surgery. This is a multidisciplinary service with surgeons from Maxillofacial and Plastic surgery backgrounds working as team members with Orthodontists, Speech and Language therapists, paediatricians and paediatric anaesthetists, audiologists and psychologists.

5. Pathways of care

In general, referrals will be received via primary or secondary care, and for trauma/burns through emergency departments.

6. Gaps and pressures

In order to comply with the proposals in Children's Surgery the infrastructure development necessary to support phased movement towards compliance would need to be the basis of a formal service review. This would require considerable revision of the way some departments work and the concentration of children's surgery onto fewer consultants would have to be reflected in their job plans. Emergency cover would be particularly difficult to staff 24/7 with European working time directives complicating things further.

Transition

Facilities for the care of adolescents need to be provided. Seamless continuing care from childhood to adult life is essential for many conditions such as cleft lip and palate. Special arrangements are necessary where treatment has been in a children's hospital.

References

1. *Children's Surgery - A First Class Service*. Royal College of Surgeons of England, 2000.
2. *National Burn Care Survey*, 2001.

UROLOGY

Table A – range of conditions suitable for management at district or PCO level

	Incidence
Foreskin problems	5% -7% males
Undescended testis	2% males
Urinary Infection	3%-5% females and males
Urinary Incontinence	5% - 10 % females and males depending on age group

1. Definition & Scope

Paediatric Urology encompasses:

- The **routine urological surgery of childhood** which can be provided by suitably trained adult urologists in District General Hospitals.
- **Specialist Paediatric Urology** provided by full time paediatric urologists or by paediatric surgeons specialising in paediatric urology, based predominantly in regional centres. Formal **supra regional specialisation** is currently limited to the management of bladder exstrophy with two NSCAG designated centres in England (Great Ormond Street London , Royal Manchester Children’s Hospital).

Table B – Uncommon conditions

- Pelvi ureteric junction obstruction ,Vesico ureteric junction obstruction (surgical and non surgical management)
- Vesico ureteric reflux (surgery)
- Duplex kidney – ureterocele
- Cystic renal malformations
- Benign or infective kidney disease
- Ambiguous genitalia, intersex
- Urinary calculi .
- Neuropathic bladder
- Urinary diversion and ‘undiversion’
- Posterior urethral valves
- Hypospadias
- Epispadias
- Investigation and management of impalpable testis
- Genito urinary malignancies in childhood
- Major urinary tract trauma
- Bladder exstrophy (including cloacal variants)- NSCAG service

2. Prevalence

Table A illustrates the conditions comprising routine, non specialised children's urology which are relatively common and can be managed by DGH adult urologists and paediatricians. Table B illustrates the conditions requiring more specialised expertise.

Voluntary organisations **include support groups for parents of children with bladder exstrophy , intersex conditions and hypospadias.**

3. Current service provision

Staffing

At PCO level staffing requirements centre principally on ensuring that surgeons (adult urologists) and anaesthetists providing the DGH service have been suitably trained , participate in continuing professional development and have sufficient workload to maintain their expertise. Likewise nurse staffing should meet national guidelines for children's nurses.

Specialist Paediatric Urological units require multidisciplinary staffing in a number of disciplines. Paediatric Urology Nurse Specialists play an invaluable role - particularly in the assessment and management of children with severe urinary incontinence.

Clinical networks are already established in many parts of the country. At present these are largely confined to the outpatient component of the service but may in future be extended to include day case surgery in DGHs.

Availability of staff

At present the majority of DGH adult urologists treat children and there is considerable scope for rationalisation to concentrate the workload in the hands of a smaller number of surgeons with a sub specialty interest in children's urology. The shortage of trainees in specialist paediatric urology is being addressed by the establishment of a dedicated training programme. Major centres will ultimately acquire the consultant establishment needed to maintain a dedicated 1 in 4 rota for paediatric urology. In smaller centres emergency cover will depend upon cross cover by general paediatric surgeons. Middle grade out of hours cover for paediatric urology is usually provided by paediatric surgical trainees (at SpR and SHO level).

4. Pathways of Care

Emergency and out of hours requirements

The emergency workload in general children's urology is low, consisting mainly of acute

scrotal pathology - notably testicular torsion, and urogenital trauma. Management of the 'acute scrotum' is best undertaken by the local on call urologist (not least because of the urgency of surgical intervention.) . The principles of trauma management are similar in adults and children . Minor trauma can be managed locally. Major trauma is rare is best managed by resuscitation and stabilisation (in collaboration with local paediatricians) followed by transfer to a regional centre. The emergency component of specialist paediatric urology is low by comparison with general paediatric surgery and consists mainly of managing acute obstruction and infection.

Travel

Wherever possible the routine elective urological surgery of childhood should be performed in DGHs by adult urologists with a subspecialty commitment to children's urology. Children with more complex conditions are referred to specialist regional units for specialised investigation and surgery but routine imaging and follow up can generally be provided on a shared basis with local DGH paediatricians and urologists. Outreach clinics undertaken by visiting Paediatric Urologists can also reduce the requirement for families to travel long distances to specialist centres.

Transition

The needs of adolescents have been under-recognised in virtually all the surgical disciplines. National guidelines have recently been proposed to redress this deficiency. Continuity of follow up into adulthood is required mainly for young people with renal insufficiency and for those who have undergone complex bladder surgery. Adolescent gynaecology is an essential component of an integrated service for individuals with intersex disorders.

5. Audit, evaluation and indicators of excellence

There has been consistent failure to fund the infrastructure and investment in information technology needed to provide systematic and credible data on surgical outcomes. Examples of good practice are almost invariably the result of initiatives established by individuals or specialty associations. The British Association of Paediatric Urologists has conducted national comparative outcome audits of pyeloplasty, posterior urethral valves and (currently) stone disease. The only NHS funded systematic audit relates to bladder exstrophy surgery in the two NSCAG-recognised units (Great Ormond Street and Manchester).

Appendices

Appendix 1: Paediatric Dentistry

Definition and scope

Paediatric Dentistry is the specialist, broad-based discipline for the oral and dental care of children.

It encompasses a variety of disciplines, techniques, procedures and skills that logically share a common basis with other dental specialties. The specialty is age based rather than technique centred. All of the dental techniques are modified, transformed or adapted to the special needs of children and adolescents. The discipline emphasises the integration of appropriate didactic and clinical skills into a framework for the comprehensive oral and dental care of children.

Table A: Children and conditions treated

<p>All Children from birth to early adulthood (generally taken as 16 years), what ever their physical, emotional, social, mental or medical condition.</p>	<p>Oral and dental disease, its prevention and treatment.</p> <p>Including:</p> <ul style="list-style-type: none"> Dental decay (caries), Gum conditions, Oral infection, Soft tissue swellings of the mouth, Malocclusion, clefting and facial dysplasia (in conjunction with Orthodontics) Developmental conditions of the teeth, including congenitally absent teeth, unerupted and buried teeth, congenitally disfigured teeth. Tooth wear. Dental injuries.
<p>Techniques used:</p> <p>Behaviour management including both pharmacological and non-pharmacological techniques.</p>	<ul style="list-style-type: none"> Soft tissue surgery Minor oral surgery Dental restorative techniques Aesthetic restorations for disfiguring conditions. Dentures for children.

Children and conditions treated

All Children from birth to early adulthood (generally taken as 16 years), what ever their physi-

cal, emotional, social, mental or medical condition. Oral and dental disease, its prevention and treatment. Including: dental decay (caries), gum conditions, oral infection, soft tissue swellings of the mouth, malocclusion, clefting and facial dysplasia (in conjunction with Orthodontics). Developmental conditions of the teeth, including congenitally absent teeth, unerupted and buried teeth, congenitally disfigured teeth. Tooth wear Dental injuries.

Techniques used: behaviour management including both pharmacological and non-pharmacological techniques. Soft tissue surgery, minor oral surgery, dental restorative techniques. Aesthetic restorations for disfiguring conditions. Dentures for children.

Incidence and Prevalence

Dental decay is widespread, however the disease is clearly linked to, and sometimes used as a measure of, social deprivation. National data are ten years old; the results of the 2003 National survey of child dental health are expected in 2004. Regional data for 2001 indicate that geographical variations in dental decay exist with a prevalence, for example, in 12-year-olds in the unfluoridated North West region, 3 times that of young people in the fluoridated West Midlands.

Debilitating medical conditions, neoplasia and its treatment, immunodeficiency, transplant and immunosuppression all may have associated oral findings. The medical care of such patients may be complicated by oral or dental disease and the need for non-elective dental treatment. The oral or dental care of these young people may be complicated by their underlying medical state.

Dental care for children with complex medical and surgical conditions may also require treatment in a specialist unit (eg issues relating to anaesthesia). 2% of children may have one or more missing teeth; between 1 in 900 and 1 in 14,000 (depending on the specific condition and particular population) will have a disfiguring dental condition

What children need

Children should be enrolled for primary care with a general dental practitioner (GDP) even before teeth are present in the mouth (6 months on average) for advice on safe weaning practices and fluoride therapy. Currently only 62 % of young people are enrolled with a GDP.

Families need speedy access to secondary and tertiary care for the less common oral conditions for infants or anxious children or children with other special needs. A fully supported, consultant provided, anaesthetic service is needed for the management of acute infection, pain and trauma, as well as for surgery or restorative care for the young, phobic or disabled child.

Voluntary Organisations

The British Society of Paediatric Dentistry, The Consultants in Paediatric Dentistry Group, The Ectodermal Dysplasia Society, CLAPA.

Current Service provision

The General Dental Services (GDS) provide primary care by individual, parent initiated registration. For those unable or unwilling to access the GDS, the community dental services (PCTs) provide complementary primary and some secondary care (e.g. sedation services).

The Hospital Dental Service, generally but not exclusively limited to the Dental Teaching Hospitals, provides tertiary care, working both alone and in conjunction with other dental or oral specialties and with paediatric colleagues.

Pathways of care

Children gain access to services through their family doctor, health visitor, general dental practitioner, community dental service, orthodontists, paediatricians.

Gaps and pressures

The vast majority of Specialists in Paediatric Dentistry were registered through a grandparenting procedure when the Specialty was recognised by the General Dental Council 5 years ago. Many of these colleagues will retire within the next ten years. Present Registrar numbers will in no way fill this void.

Consultants in Paediatric Dentistry (who are also Specialists) are predominantly based in the Dental Teaching Hospitals. This leads to large geographical areas of the country being without a tertiary service. There is a real role for them to be both Community- and Children's Hospital-based as well.

A fully supported, consultant led, anaesthetic service is not equitably present across the United Kingdom and some children may wait many weeks in acute pain for such dental care.

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