Received: 8 April 2024 | Accepted: 2 July 2024

DOI: 10.1002/jpn3.12319

JPGN

ORIGINAL ARTICLE

Hepatology

Assessment of disease severity with magnetic resonance cholangiography in pediatric-onset primary sclerosing cholangitis

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Funding information

This study was funded by Helsinki University Hospital Diagnostic Center, Päivikki and Sakari Sohlberg Foundation, Abdominal Radiologists of Finland, and Pediatric Research Foundation

Abstract

Objectives: Magnetic resonance cholangiopancreaticography (MRCP) has supplanted endoscopic retrograde cholangiopancreaticography (ERCP) as the preferred imaging modality for primary sclerosing cholangitis (PSC). However, data about the accuracy of MRCP in assessing disease severity are limited, particularly in children. We assessed the accuracy of MRCP in disease severity evaluation and investigated the correlation between imaging findings and biochemical parameters (including the multivariate risk index SCOPE) in patients with pediatric-onset PSC.

Methods: We included 36 patients with PSC (median age: 16) who had MRCP and ERCP performed within 4-month intervals. Two experts, blinded to ERCP findings, evaluated the bile duct changes in consensus using the Modified Amsterdam PSC Score. The agreement between MRCP and ERCP evaluations was tested with weighted kappa statistics and the correlation between disease severity and biochemical parameters with Spearman's rank correlation.

Results: The agreement between MRCP and ERCP was good for extrahepatic (weighted kappa 0.69; 95% CI 0.53-0.84) but fair for intrahepatic (weighted kappa 0.35; 95% CI 0.14-0.56) bile ducts. Intrahepatic and extrahepatic MRCP scores correlated with APRI ($\rho = 0.42$, $p = 0.020$ and $\rho = 0.39$, $p = 0.033$, respectively), while extrahepatic MRCP score also correlated with biliary neutrophils ($\rho = 0.36$, $p = 0.035$). We found a good correlation between the SCOPE index and intrahepatic MRCP score ($\rho = 0.53$, $p = 0.004$), and extrahepatic MRCP score ($\rho = 0.57$, $p = 0.001$).

Conclusions: MRCP is accurate at evaluating the severity of extrahepatic bile duct changes in pediatric-onset PSC but tends to underestimate intrahepatic changes. The SCOPE index's robust correlation with imaging scores supports its role as a comprehensive diagnostic tool, outperforming individual laboratory metrics.

KEYWORDS

bile duct, children, liver

1 | INTRODUCTION

Primary sclerosing cholangitis (PSC) is a chronic liver disease characterized by biliary inflammation and fibrosis that lead to strictures of intra- and extrahepatic bile ducts. The natural history of PSC is progression to biliary cirrhosis and, eventually, end-stage liver disease.[1] PSC is also a preneoplastic condition predisposing to hepatobiliary and colorectal cancers, especially in adults.[1–4] Although PSC is often a milder disease in children, almost half of the patients with pediatric-onset PSC develop an adverse liver outcome after 10 years.[5] Currently, no medical therapy can prevent PSC progression, and liver transplantation is considered with advanced disease.[3] To allow timely liver transplantation, imaging is crucial for the early detection of complications.

Previously, endoscopic retrograde cholangiopancreaticography (ERCP) was the gold standard imaging method in PSC.[6,7] Due to the relatively high complication rate in ERCP, noninvasive and safe magnetic resonance cholangiopancreaticography (MRCP) has largely replaced ERCP as the primary imaging method.[8,9] ERCP has, however, proven better at detecting early PSC.[10–12] Besides this, literature on the comparison of MRCP and ERCP in disease severity evaluation is scarce. Although MRCP has proven reliable in diagnosing PSC,[11,13–15] only a few studies have compared MRCP and ERCP in disease severity assessment,[16–18] with only one such study conducted on pediatric patients.[17] Furthermore, there is little published research on the correlation between the severity of cholangiographic findings and biochemical parameters.[18–21] Thus, the aims of this study were (1) to evaluate the agreement of MRCP and ERCP in the assessment of disease severity, and (2) to evaluate the correlation of the cholangiographic disease severity and biochemical parameters in patients with pediatric-onset PSC.

What is Known

- Imaging plays a pivotal role in the diagnosis, monitoring, and evaluation of complications in primary sclerosing cholangitis (PSC).
- While magnetic resonance cholangiopancreatography (MRCP) reliably diagnoses PSC, its role in assessing disease severity is uncertain.

What is New

- MRCP is accurate at evaluating the severity of extrahepatic bile duct changes in pediatric-onset PSC but tends to underestimate intrahepatic changes.
- The SCOPE index exhibited a moderate to strong correlation with MRCP and ERCP scores, suggesting it may serve as a potential indicator of PSC disease severity.

2 | METHODS

2.1 | Study design, time, and population

This retrospective cohort study was performed at Helsinki University Hospital, Finland, a tertiary-care hospital. We identified patients with pediatric-onset PSC from the national PSC registry founded in 2010. This registry contains clinical data and biochemical, histological, and cholangiographic findings of patients with PSC. Patients with secondary sclerosing cholangitis and IgG4-associated cholangitis were not included. The diagnostic criteria for PSC followed the EASL guidelines.[9] At the time of the study, ERCP was performed at our center for all patients with PSC to confirm the diagnosis, assess disease progression, evaluate biliary tract dysplasia, and determine the need for endoscopic treatment.[22]

From the PSC registry, we found 61 patients diagnosed with PSC at the age of 4–18 years between 1999 and 2017. Of the 61 patients, we included those who were imaged with both MRCP and ERCP within a 4-month interval between the age of 4-25 years. The 4-month interval was chosen to reduce bias due to disease progression between these two studies. We only included study pairs with MRCP performed before ERCP so that the possible endoscopic therapy (i.e., dilatation or stenting) did not affect the stricture appearance in MRCP. For patients with several MRCP/ERCP -study pairs, we selected the most recent pair to include studies performed with the most modern MR devices. We only included examinations carried out before liver transplantation and MRCP:s performed with 1.5/3 Tesla magnetic resonance unit. The patient flowchart is shown in Figure 1.

2.2 | MRCP

MRCP examinations were acquired with torso phased-array coils on 1.5 T MR unit (Siemens Avanto [$n = 13$], Sonata [$n = 1$] or Symphony [$n = 6$], Siemens Healthcare, Erlangen, Germany; GE Optima [$n = 2$], GE Medical Systems, Milwaukee, WI; Philips Achieva [$n = 13$], Philips Medical Systems, Best, Netherlands; Toshiba Titan [$n = 1$], Canon Medical Systems, Tustin, California). Patients were advised to fast for 3 h before examination. Before imaging, patients drank pineapple juice to diminish the signal of bowel contents and to better visualize the biliary tree.

The imaging protocol included the following sequences: axial and coronal T2-weighted sequences, axial T2-weighted sequence with fat suppression, axial in and out of phase T1-weighted gradient echo sequence, respiratory triggered heavily T2-weighted 3D sequence, maximum intensity projection (MIP) images reconstructed from the 3D sequence, and breath-hold thick-slab single-shot heavily T2-weighted sequence in coronal and oblique coronal planes.

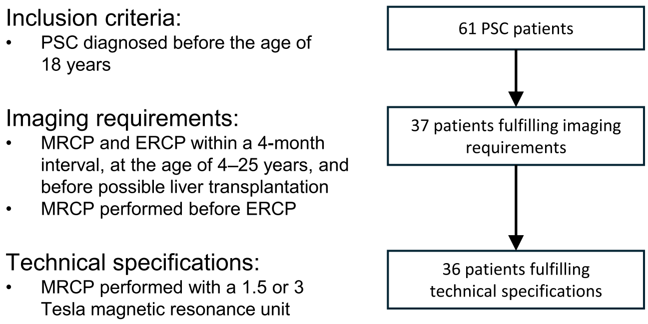

FIGURE 1 Patient inclusion flowchart. Modified from Vanhanen et al.[23] PSC=primary sclerosing cholangitis, MRCP= magnetic resonance cholangiopancreaticography, ERCP= endoscopic retrograde cholangiopancreaticography.

An experienced abdominal radiologist (A.O.) and a radiologist specializing in abdominal radiology (E.V.) retrospectively analyzed the MRI images in consensus, blinded to the ERCP evaluations and clinical information.

2.3 | ERCP

ERCP studies were performed by experienced endoscopists at Helsinki University Hospital, Finland. A previous study from our center describes the ERCP protocol in detail.[18] ERCP images were retrospectively reviewed by an experienced endoscopist (A.T.) blinded to MRCP evaluations and clinical information.

2.4 | Image analysis and definitions

Bile duct changes detected in MRCP and ERCP were evaluated according to a modified version of Amsterdam PSC Score (Supplemental digital content, Table S1).[18] Amsterdam PSC Score was originally described in 2001 by Rajaram and colleagues and scores intra- and extrahepatic bile ducts based on the severity of PSC-associated changes.[24] We used a modified version of the score, where intrahepatic score 1 is separated into two categories (i.e., 1 = ductular irregularities, 2 = multiple caliber changes, minimal dilatation) to enable more precise classification of early changes. Although developed for ERCP, this score was also applied to MRCP images to objectively compare the biliary findings. The MRCP and ERCP reviewers scored separately the following anatomical sites: (1) right hepatic duct and right lobe's intrahepatic ducts, (2) left hepatic duct and left lobe's intrahepatic ducts, (3) common hepatic duct and (4) common bile duct.

2.5 | Biochemical parameters

The following biliary parameters were evaluated: (1) biliary neutrophils (0 = absent, 1 = mild, 2 = high), and Papanicolaou cytology classification (1 = normal epithelium, 2 = benign atypia, 3 = mild suspicion of neoplasia, cytologically corresponding to low-grade dysplasia, 4 = high suspicion of neoplasia, cytologically corresponding to high-grade dysplasia, 5 = malignancy, cytologically corresponding to carcinoma).

In addition to biliary parameters, the following plasma/serum parameters were included as surrogate markers of disease activity and severity: gamma-glutamyl transpeptidase (GGT), alkaline phosphatase (ALP), alanine aminotransferase (ALT), total bilirubin, carbohydrate antigen 19-9 (CA19-9), aspartate aminotransferase-to-platelet ratio index (APRI). The biliary markers were evaluated from bile samples gathered during the ERCP included in the analysis, and the plasma/serum markers from blood samples obtained closely with MRCP/ERCP imaging. We also correlated MRCP and ERCP scores with the SCOPE index, a validated pediatric-specific prognostic tool that includes multiple laboratory studies (total bilirubin, albumin, platelet count, gamma glutamyltransferase) and cholangiography.[25]

2.6 | Statistical analysis

For the analyses, a mean score of intrahepatic ducts (right and left lobe's intrahepatic ducts) and extrahepatic ducts (common hepatic duct and common bile duct) was used. The agreement between MRCP and ERCP evaluations was tested with weighted kappa statistics using quadratic weights. For this, we grouped the mean scores into three categories (0 = no changes, 1-2 = mild changes, and 3-4 = severe changes), as described in a previous study conducted on adult patients in our center.[18] The relationships between MRCP and ERCP Modified Amsterdam scores and biochemical parameters were assessed with Spearman's rank correlation. $p < 0.05$ was considered significant. Analyses were performed with IBM SPSS Statistics for Windows, Version 25.0. Armonk, NY: IBM Corp.

2.7 | Ethical approval information

According to Finnish legislation, ethical approval or informed consent were not required as the patients were not contacted in this register-based study. The study protocol (number 64/13/03/03/2012) was approved by the local Ethics Committee of Helsinki University Hospital.

3 | RESULTS

3.1 | Patient characteristics

In total, ERCP-MRCP pairs of 36 patients with pediatric-onset PSC were included in the analyses. Characteristics of the patients are shown in Table 1.

3.2 | Agreement between MRCP and ERCP Modified Amsterdam scores

In ERCP, 35/36 patients (97%) had intrahepatic duct changes, and 16/36 patients (44%) had extrahepatic duct changes. The sensitivity of MRCP in detecting any PSC changes was 89% (31/35) for intrahepatic ducts and 94% (15/16) for extrahepatic ducts. In scoring disease severity, the agreement between MRCP and ERCP was only fair for intrahepatic bile ducts (weighted kappa 0.35; 95% CI 0.14–0.56), and MRCP tended to score the changes as less severe than ERCP (Figure 2). For extrahepatic bile ducts, the agreement between MRCP and ERCP scores was good (weighted kappa 0.69; 95% CI 0.53–0.84). In six cases (17%), ERCP found no changes in extrahepatic bile ducts, but MRCP revealed mild changes. In contrast, in seven cases (19%) where ERCP detected severe changes, MRCP classified the changes as mild. The results are shown in Table 2. ERCP-related complication occurred in 4/36 patients (11%), three cases being mild pancreatitis and one guide-wire perforation. At the time of the study, one patient had been diagnosed with cirrhosis.

TABLE 1 Patient characteristics.

Patients	36
Female	14 (39%)
Age at PSC diagnosis, years	15.3 (12.0–16.5)
Age at MRCP, years	16.0 (14.5–19.6)
Interval between MRCP and ERCP examinations, months	2.5 (1.5–3.4)
PSC with features of autoimmune hepatitis	13 (36%)
Inflammatory bowel disease	32 (89%)

Note. Continuous variables are expressed as medians with interquartile ranges. Modified from Vanhanen et al.[23]

Abbreviations: PSC, primary sclerosing cholangitis; MRCP, magnetic resonance cholangiopancreaticography; ERCP, endoscopic retrograde cholangiopancreaticography.

3.3 | Modified Amsterdam scores and biochemical parameters

Intrahepatic MRCP score correlated moderately with APRI ($\rho = 0.42$, $p = 0.020$), but not with other parameters. Extrahepatic MRCP score correlated with APRI ($\rho = 0.39$, $p = 0.033$) and biliary neutrophils ($\rho = 0.36$, $p = 0.035$). Median values grouped by MRCP scores are shown in Supplemental digital content, Table S2.

Intrahepatic ERCP score correlated with ALP ($\rho = 0.50$, $p = 0.003$), ALT ($\rho = 0.35$, $p = 0.045$), APRI ($\rho = 0.42$, $p = 0.021$), and biliary neutrophils ($\rho = 0.39$, $p = 0.022$), but not with other parameters (GGT, bilirubin, CA19-9, and biliary cytology). Extrahepatic ERCP score correlated with biliary neutrophils ($\rho = 0.50$, $p = 0.002$), and biliary cytology ($\rho = 0.38$, $p = 0.029$). Median values grouped by ERCP scores are shown in Supplemental digital content, Table S3.

The data needed to calculate the SCOPE index was available for 28 patients (78%). We found a moderate to strong correlation between the SCOPE index and intrahepatic MRCP score ($\rho = 0.53$, $p = 0.004$), extrahepatic MRCP score ($\rho = 0.57$, $p = 0.001$), intrahepatic ERCP score ($\rho = 0.63$, $p = <0.001$), and extrahepatic ERCP score ($\rho = 0.54$, $p = 0.003$).

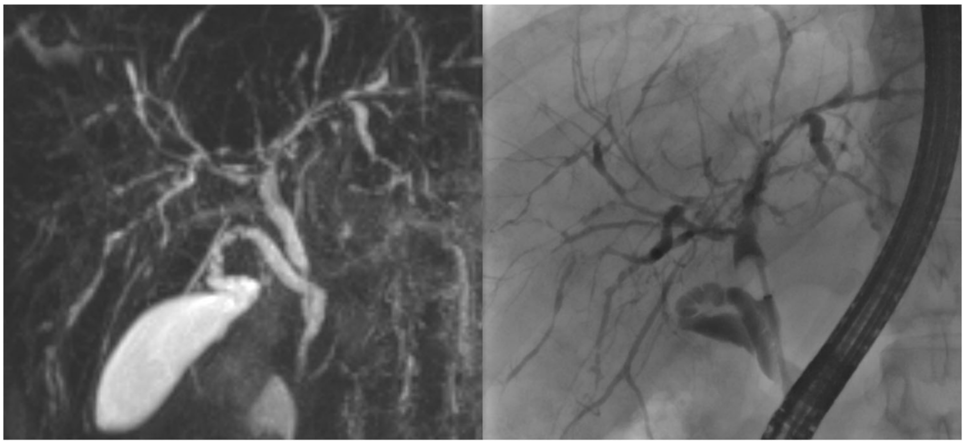

FIGURE 2 An example of incongruent grading in magnetic resonance cholangiopancreaticography (MRCP) and endoscopic retrograde cholangiopancreaticography (ERCP). In MRCP, intrahepatic changes were graded 1 in the right hepatic duct and right lobe's intrahepatic ducts, and grade 2 in the left hepatic duct and left lobe's intrahepatic ducts. In ERCP, ductal changes are more prominent and were graded 3 in both lobes.

TABLE 2 Agreement between magnetic resonance cholangiography (MRCP) and endoscopic retrograde cholangiography (ERCP) in assessing cholangiographic changes of primary sclerosing cholangitis (PSC) using Modified Amsterdam score.

Intrahepatic ducts

	ERCP score		
MRCP score	0 ($n = 1$)	1–2 ($n = 21$)	3–4 ($n = 14$)
0	1	4	0
1–2	0	16	11
3–4	0	1	3

Extrahepatic ducts

	ERCP score		
MRCP score	0 ($n = 20$)	1–2 ($n = 5$)	3–4 ($n = 11$)
0	14	1	0
1–2	6	4	7
3–4	0	0	4

4 | DISCUSSION

In this study assessing MRCP and ERCP in disease severity evaluation of pediatric-onset PSC, the agreement between MRCP and ERCP was only fair for intrahepatic bile ducts but good for extrahepatic bile ducts. The SCOPE index exhibited a good correlation with MRCP and ERCP scores, suggesting it may serve as a potential indicator of PSC disease severity. Of the single laboratory parameters, APRI correlated with intrahepatic MRCP and ERCP scores, as well as extrahepatic MRCP score.

4.1 | Agreement between MRCP and ERCP Modified Amsterdam scores

Previously, a few studies have compared MRCP and ERCP in PSC disease severity evaluation. Two studies found a good correlation between MRCP and ERCP findings, but the study population was small in both studies (nine adult PSC patients and 21 pediatric patients).[16,17] In contrast, a previous study on 48 adult PSC patients in our center revealed only moderate agreement between MRCP and ERCP, especially for intrahepatic ducts.[18]

According to this study, MRCP tends to score intrahepatic changes as less severe than ERCP. MRCP ignored four cases with mild intrahepatic changes, which is consistent with previous studies indicating that ERCP is better at detecting early PSC changes.[10–12] This difference is probably explained by the poorer spatial resolution of MRCP as compared to ERCP. Furthermore, MRCP tended to underestimate severe intrahepatic changes. In a previous study on adult PSC patients,[18] MRCP also underestimated severe biliary changes in 19% of patients. In that study, however, MRCP also overestimated moderate intrahepatic ductal changes in 39% of the patients. This occurred in only one patient with moderate intrahepatic duct changes in the current study.

For extrahepatic ducts, we found substantial agreement between MRCP and ERCP. The incongruent evaluations between MRCP and ERCP included both false negative and false positive observations. On one hand, MRCP tended to underestimate severe changes noticed by ERCP, as also seen with intrahepatic changes. On the other hand, MRCP showed mild changes in six ERCP-negative cases (6/20, 30%). In a previous study on adult patients, MRCP tended to overestimate extrahepatic ductal changes.[18] The reason for these false positives in MRCP may be the lack of filling pressure used in ERCP, as shown in our previous study.[23] While MRCP is a static method, the pressure injection of contrast media allows the evaluation of the distensibility of the strictures in ERCP.

Although previously regarded as the gold standard imaging method in PSC, ERCP also has certain limitations. In addition to its invasive nature and the risk of ERCP-related complications,[26] ERCP may fail to visualize bile ducts above tight strictures, leading to false negative findings.[14,16,20,27,28] It is also important to acknowledge that even with structured grading system, the evaluation of subtle changes in small structures is prone to subjective interpretation, particularly when relying on descriptive criteria. In a study assessing interobserver agreement of MRCP and ERCP in PSC, agreement was poor for both MRCP and ERCP when disease severity was evaluated.[28] That study did not, however, compare MRCP with ERCP.

As Amsterdam score is developed for ERCP, it may not be ideal for grading MRCP, although often used in studies comparing MRCP and ERCP. In a study by Ernst et al. a minor modification was made for MRCP assessment of severe intrahepatic involvement: instead of "severe pruning" applied to ERCP, severe involvement was considered when long strictures were seen without connection between central and peripheral bile ducts.[16] Although Amsterdam score is not in clinical use for MRCP grading, recent guidelines for reporting MRCP/MRI findings in PSC patients encompass similar elements.[29]

In addition to stricture assessment, MRCP provides information on the background liver changes, such as development of portal hypertension and cirrhosis, enhancing the overall value of MRCP. In this study, we did not assess liver parenchymal changes. As the superior safety profile of MRCP has established it as the primary imaging method in PSC, future studies are needed to investigate the predictive value of MRCP and liver MRI findings in PSC, especially in pediatric patients.

4.2 | Modified Amsterdam scores and biochemical parameters

We performed an exploratory analysis on the MRCP and ERCP scores and clinical markers of disease activity. According to our results, APRI correlated with intra- and extrahepatic MRCP scores as well as intrahepatic ERCP score. To our knowledge, the correlation between APRI and severity of biliary changes has not been studied before. Instead, APRI associated with a poor prognosis in a multicenter study evaluating pediatric PSC.[5] In adults with PSC, APRI correlated with advanced disease defined in histopathological examination.[30] It has been suggested that APRI may serve as an indicator of hepatic fibrosis while bilirubin and GGT may be markers of cholangiopathy in PSC.[5]

Several studies on adults with PSC have reported the prognostic value of ALP.[31–33] In a previous study on adult patients in our center, intrahepatic MRCP and ERCP scores had a statistically significant association with ALP.[18] In children, however, ALP is generally considered unreliable due to the impact of bone growth on ALP isozymes. Instead of ALP, GGT is recommended as the biochemical marker of biliary disease in pediatric patients.[1,5] In our study, however, GGT did not correlate with cholangiographic changes. Instead, we found a moderate correlation between intrahepatic ERCP Modified Amsterdam score and ALP. As the median age of our patients was 16, it is possible that the effect of growth on ALP was not significant anymore. Also, the relatively small sample size may have influenced the results. Finally, cholestatic liver enzymes might fluctuate during the course of the disease, not necessarily reflecting a more severe disease.[22] In two previous studies on children with PSC, intra- and extrahepatic MRCP scores did not correlate with ALP or GGT.[21,34]

Intra- and extrahepatic ERCP scores correlated with the level of biliary neutrophils, and biliary cytology correlated with extrahepatic ERCP score. Unexpectedly, MRCP scores did not show such correlation with biliary cytology, but extrahepatic score and biliary neutrophils were correlated. We did not find correlation between MRCP and ERCP scores and Ca19-9, but only two of our patients (5.6%) had cytology suggestive of malignancy (Papanicolaou stage 3) at the time of study, and none was diagnosed with cancer. These findings are contrary to the study on adult patients with PSC, where MRCP and ERCP Modified Amsterdam scores did not associate with biliary neutrophils or cytology, but Ca19-9 was associated with intrahepatic MRCP and ERCP score, as well as extrahepatic MRCP score.[18] Due to the exploratory nature of both studies, however, these findings should be interpreted with caution.

The SCOPE index demonstrated a stronger correlation with the MRCP and ERCP scores compared to any individual laboratory value. It is plausible that the prognostic significance of a single laboratory value in PSC is limited, and the observed higher correlation with the SCOPE index underscores the potential advantage of a composite risk index like SCOPE.[25] It is noteworthy, however, that the SCOPE index's positive correlation with ERCP and MRCP may be partly influenced by its inclusion of the bile duct status as one of its components.

4.3 | Strengths and limitations

ERCPs were performed in Helsinki University Hospital according to a standard protocol by experienced endoscopists. Two abdominal radiologists evaluated MRCPs; however, due to consensus reading, interrater agreement was not calculated. To minimize the bias due to disease progression between MRCP- and ERCP -studies, we only included imaging studies performed within a 4-month interval. The study population consisted of patients with pediatric-onset PSC, a target population with little previous published research. The limitations of this study include retrospective design, and the limited number of included patients. However, given the rarity of PSC, particularly in children, the study population was considered adequate and is comparable to previous research on pediatric PSC. The nature of the correlation analyses involving Modified Amsterdam scores and clinical markers of disease activity was explorative.

4.4 | Conclusions

MRCP is accurate at evaluating extrahepatic bile duct changes but tends to underestimate the severity of intrahepatic changes in pediatric-onset PSC. The SCOPE index exhibited a moderate to strong correlation with MRCP and ERCP scores, surpassing the predictive capability of individual laboratory parameters. Among the single parameters, APRI showed a significant correlation with intrahepatic MRCP and ERCP scores, as well as with the extrahepatic MRCP score.

ACKNOWLEDGMENTS

RN Anne Nikkonen is thanked for her help in data collection. This study was funded by Helsinki University Hospital Diagnostic Center, Päivikki and Sakari Sohlberg Foundation, Abdominal Radiologists of Finland, and Pediatric Research Foundation. The funders had no role in the design and conduct of the study; collection, management, analysis, and interpretation

of the data; preparation, review, or approval of the manuscript; and decision to submit the manuscript for publication.

CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

How to cite this article: Vanhanen E, Ovissi A, Kolho K-L, Tenca A. Assessment of disease severity with magnetic resonance cholangiography in pediatric-onset primary sclerosing cholangitis. *J Pediatr Gastroenterol Nutr*. 2024;1-8. doi:10.1002/jpn3.12319