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Magnetic resonance cholangiopancreatography as a diagnostic tools to diagnose biliary atresia at Dr. Soetomo hospital

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Abstract

Background: Cholestatic jaundice results from diminished bile flow and/or excretion, and can be caused by a number of disorders such as biliary atresia (BA). Magnetic resonance cholangiopancreatography (MRCP) is widely accepted as one of modalities for biliary system imaging, but liver biopsy still generally used for BA diagnosis, especially in developing countries. **Objective:** To evaluate diagnostic value of biliary atresia from MRCP to the result of liver biopsy. **Methods:** A cross sectional for diagnostic study documented of hospitalized patients from June 2014 to June 2015. All patients were had MRCP and liver biopsy examination. Data of age, gender, clinical manifestation and result of MRCP and liver biopsy with ROC to evaluate the sensitivity and specificity was done. Liver biopsy revealed of biliary atresia was made based on proliferation, degeneration and fibrosis of bile ducts. ROC to evaluate the sensitivity and specificity was done. The sensitivity, specificity, negative predictive value, positive predictive value of MRCP in diagnosing BA were calculated. **Results:** There were 16 patients enrolled in this study with median age of diagnose was 6(range 3-11) months old. There were 9/16 girl. The median age of onset of jaundice was 5(range 2-14) days. All patients had hepatomegaly and splenomegaly. Histopathology from liver biopsy revealed biliary atresia in 12/16 patients. From ROC curve, sensitivity of MRCP was 87.5% and specificity 62.5% with PPV 70% and NPV 80%. Five patients underwent of Kasai procedure and revealed biliary atresia. **Conclusion:** MRCP is sensitive but not specific for diagnosing BA and MRCP has moderate sensitivity and specificity for BA diagnosis. **Keyword:** *biliary atresia, magnetic resonance cholangiopancreatography, liver biopsy, diagnostic test.*

ABSTRAK

Latar belakang: Ikterus dikarenakan kolestasis terjadi akibat berkurangnya aliran empedu dan / atau ekskresi, dan dapat disebabkan oleh sejumlah gangguan seperti atresia biliaris (BA). Magnetic resonance cholangiopancreatography (MRCP) secara luas diterima sebagai salah satu modalitas untuk pencitraan sistem empedu, tetapi biopsi hati masih secara umum digunakan untuk diagnosis BA, terutama di negara berkembang. **Tujuan:** Untuk mengevaluasi nilai diagnostik atresia biliaris dari MRCP ke hasil biopsi hati. **Metode:** Sebuah cross sectional untuk studi diagnostik didokumentasikan pasien rawat inap dari Juni 2014 hingga Juni 2015. Semua pasien menjalani MRCP dan pemeriksaan biopsi hati. Data usia, jenis kelamin, manifestasi klinis dan hasil MRCP dan biopsi hati dengan ROC untuk mengevaluasi sensitivitas dan spesifisitas dilakukan. Biopsi hati mengungkapkan atresia biliar dibuat berdasarkan proliferasi, degenerasi dan fibrosis saluran empedu. ROC untuk mengevaluasi sensitivitas dan spesifisitas yang dilakukan. Sensitivitas, spesifisitas, nilai prediktif negatif, nilai prediksi positif MRCP dalam mendiagnosis BA dihitung. **Hasil:** Ada 16 pasien yang

terdaftar dalam penelitian ini dengan median usia diagnosa adalah 6 (kisaran 3-11) bulan. Ada 9/16 gadis. Usia rata-rata onset penyakit kuning adalah 5 (kisaran 2-14) hari. Semua pasien mengalami hepatomegali dan splenomegali. Histopatologi dari biopsi hati mengungkapkan atresia bilier pada 12/16 pasien. Dari kurva ROC, sensitivitas MRCP adalah 87,5% dan spesifisitas 62,5% dengan PPV 70% dan NPV 80%. Lima pasien menjalani prosedur Kasai dan mengungkapkan atresia bilier. **Kesimpulan:** MRCP sensitif tetapi tidak spesifik untuk mendiagnosis BA dan MRCP memiliki sensitivitas dan spesifisitas sedang untuk diagnosis BA.

Kata kunci: *biliary atresia, magnetic resonance cholangiopancreatography, liver biopsy, diagnostic test.*

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INTRODUCTION

Cholestatic jaundice results from diminished bile flow and/or excretion, and can be caused by a number of disorders such as (BA) biliary atresia (Moyer V et al,2014) BA is a devastating disease of infancy where the bile ducts are occluded and destroyed by a fibroinflammatory process. BA is rare but still the most common indication for childhood liver transplantation (LT). BA treatment is started with a Kasai Procedure or portoenterostomy (PE) operation, adjuvant medical therapy, and continued with LT if the PE fails. (Liu Bo, et al, 2014) Magnetic resonance cholangiopancreatography (MRCP) is widely accepted as one of modalities for biliary system imaging, but liver biopsy still generally used for BA diagnosis, especially in developing countries. This present study was to evaluate diagnostic value of biliary atresia from MRCP to the result of liver biopsy.

SUBJECTS AND METHODS

This cross sectional study included children admitted in pediatric wards Soetomo Hospital with suspected BA during June 2014 to June 2015. Inclusion criteria was 16 patients with consecutive cholestatic infants (9 girls, 7 boys) with a final diagnosis of INH or BA were entered in our study as clinical condition characterized by jaundice and all patients were had MRCP and liver biopsy examination. Data of age, gender, clinical manifestation and result of MRCP and liver biopsy. Liver biopsy revealed of biliary atresia was made based on proliferation, degeneration and fibrosis of bile ducts. The sensitivity, specificity, negative predictive value, positive predictive value of MRCP in diagnosing BA were calculated.

RESULTS

Clinical characteristics of patients are summarized in table 1. There were 16 patients enrolled in this study with median age of diagnose was 6 (range 3-11) months old. There were 9/16 girl. The median age of onset of jaundice was 5 (range 2-14) days. All patients had hepatomegaly and splenomegaly. Histopathology from liver biopsy revealed biliary atresia in 12/16 patients.

Table 1. Patient characteristic children with suspected Biliary Atresia admitted in pediatric ward 2014-2015.

Characteristics	Value
Sex	
• Male	7 (43.8)
• Female	9 (56.2)
Age	
• < 3 month	2 (12.5)
• > 3 month	14 (87.5)
Dark Urine	
• Yes	13 (81.2)
• No	3(18.8)
Acholis Stool	
• Yes	14 (87.5)
• No	2(12.5)
Splenomegaly	
• Yes	15 (93.8)
• No	1 (6.2)
Hepatomegaly	16(100.0)

From table 2x2 (table2), sensitivity of MRCP was 87.5% and specificity 62.5% with positive predictive value (PPV) 70% and negative predictive value (NPV) 80%. Five from 16 patients underwent of Kasai procedure and revealed biliary atresia.

Table 2. Analysis MRCP compare with liver biopsy in BA patients.

MRCP +/-	Liver Biopsy +/-			
		+	-	Total
+	+	7	3	10
-	+	1	5	6
		8	8	16

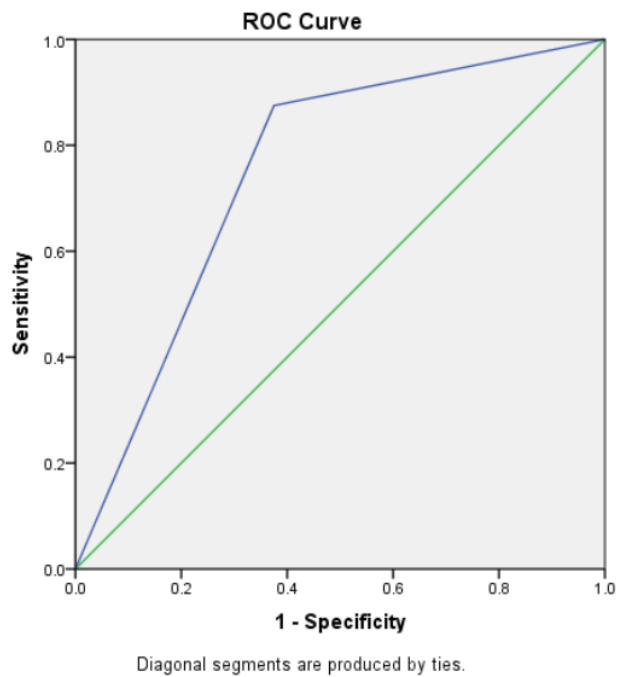


Figure 1. ROC MRCP compare with liver biopsy in BA patient

DISCUSSION

The North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) guideline for the evaluation of cholestatic jaundice in infants recommends that any infant noted to be jaundiced at the 2-week well-child visit should be evaluated for cholestasis (Moyer V, et al, 2014) Evaluation of breast-fed infants may be delayed until 3 weeks of age if they have a normal physical examination, no history of

dark urine or light stools, and can be reliably monitored (Liu Bo, et al, 2011) Neonatal hepatitis and BA, which typically occur in term infants, account for 70-80% of cases(Hartley,2009)

In this study liver biopsy revealed of billiary atresia was made based on proliferation, degeneration and fibrosis of bile ducts. Percutaneous liver biopsy is used by most centres in the differential diagnostic path of a cholestatic infant. The liver histological assessment has approximately 90-100% sensitivity and 80-98% specificity for biliary obstruction.(Takaya, 2007) Previous study revealed liver biopsy of cholestatic infants, the features indicating BA were bile plugs in bile ducts and canaliculi, portal tract edema, severe portal fibrosis, and bile duct proliferation, whereas sinusoidal fibrosis ruled against BA(Yang Ji, 2009) Considerable interobserver variability was observed: the percentage of agreement in the different features assessed varied from 43% to 93%.(Khalil BA, 2010) Percutaneous liver biopsy is generally employed in the evaluation of neonatal cholestasis, particularly when biliary tract obstruction is high on the differential diagnosis (Humprey TM, 2007) The biopsy should be interpreted by a pathologist with expertise in pediatric liver disease. Biopsy is recommended before performing a surgical procedure to diagnose BA. (Norton KI, et.al, 2012)

A promising new option for imaging infants in whom biliary atresia is suspected with magnetic resonance cholangiopancreatography (MRCP). MRCP can provide clear images of intrahepatic ducts and the common bile duct. The delineation of an incomplete extrahepatic bile duct and atrophic gall bladder on MR cholangiography suggests biliary atresia. Liver biopsy may help in the diagnosis of biliary atresia. (Metreweli C, 2004)

In our study, MRCP had a sensitivity and a specificity of 87.5 % and 62.5%, PPV 70% and NPV 80% for BA. We found that MRCP had higher sensitivity and NPV in the identification of BA, but the specificity and PPV were lower. This lower accuracy may be due to the lower experience of our sonographer for detection of triangular cord sign. Contrary to previous reports, false-positive and false-negative findings occur at MRCP. (Liu Bo, et al, 2012)

Conclusion

MRCP has moderate sensitivity and specificity for BA diagnosis

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