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## INSTRUCTION TO AUTHOR
Complications of Biliary Atresia in a 27-Year-Old Male Patient

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ABSTRACT

Biliary atresia (BA) is a disease of the extrahepatic biliary tree that presents with biliary obstruction in the neonatal period, which is caused by fibro-obliterative process. Kasai procedure, a hepatopanenterostomy (HPE) as an attempt to restore bile flow from the liver to the proximal small bowel, has been shown to improve survival in BA patients. Many BA survivors who had undergone Kasai HPE will have slowly progressive liver disease and the majority of patients will ultimately require liver transplantation. In spite of many experimental treatments, cirrhosis still occurs in BA patients survival.

This case report presents a male patient with biliary atresia that has survived for 27 years after Kasai procedure. He had been repeatedly admitted to hospital with complications caused by cirrhosis, such as repeated variceal and hemorrhoid bleeding and also refractory ascites. These complications are indications for liver transplantation. Although Kasai HPE procedure improves survival in BA patients in Indonesia, long-term complications of cirrhosis makes the patient awaits for liver transplantation.

Keywords: biliary atresia, Kasai procedure, hepatopanenterostomy, cirrhosis, liver transplantation

ABSTRAK

Atresia bilier (AB) adalah penyakit pada saluran bilier ekstrahepatik yang menimbulkan sumbatan bilier pada periode neonatal disebabkan proses fibro-obliterasi. Prosedur Kasai merupakan suatu hepatopanenterostomi (HPE) adalah tindakan untuk mengalihkan aliran empedu dari hati ke usus kecil, telah meningkatkan kesuksesan pasien AB. Kebanyakan pasien AB yang selamat setelah menjalani Kasai HPE akan menderita penyakit hati yang progresif dan akhirnya mayoritas pasien akan membutuhkan transplantasi hati. Walaupun terdapat banyak terapi yang telah dicoba diberikan, sirosis tetap terjadi pada pasien AB yang selamat.

Kasus ini melaporkan pasien laki-laki dengan atresia bilier yang dapat bertahan sampai 27 tahun setelah menjalani prosedur Kasai dan mendapat perawatan berulang kali karena komplikasi sirosis seperti perdarahan varises dan hemoroid serta refraksi astites. Komplikasi yang terjadi merupakan indikasi dari transplantasi hati. Walau pun prosedur Kasai HPE dapat meningkatkan kesuksesan pasien AB di Indonesia, komplikasi jangka panjang seperti sirosis masih memerlukan penatalaksanaan lanjutan yaitu transplantasi hati.

Kata kunci: atresia bilier, prosedur Kasai, hepatopanenterostomi, sirosis, transplantasi hati

INTRODUCTION

Biliary atresia (BA) is a progressive, idiopathic, fibro-obliterative disease of the extrahepatic biliary tree that presents with biliary obstruction exclusively in the neonatal period. The overall incidence of BA is low, i.e. about one in 10,000 to 20,000 live births. The incidence of BA varies from 5/100,000 live births in The Netherlands, 6/100,000 in the British Isles,