

Unraveling a complex case of Biliary Atresia Splenic Malformation Syndrome (BASM): A case report

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Abstract

Introduction:

Biliary atresia is a congenital obstruction of the bile ducts that can result in progressive liver damage (cirrhosis and fibrosis) and requires early surgical intervention.

Case Presentation: This case report discusses a unique presentation of biliary atresia that presented with proximal intestinal obstruction in the form of duodenal atresia, a preduodenal portal vein, malrotation, with a normal spleen and no associated congenital cardiac anomalies. It highlights the diagnostic challenges and management strategies applied in syndromic biliary atresia.

Conclusion:

The below case demonstrates a rare presentation of syndromic biliary atresia and the importance of early surgical intervention.

Keywords: Syndromic biliary atresia, splenic malformation, congenital, Kasai portoenterostomy

