

Case Report

Anti-Reflux Valve Procedure in Recurrent Ascending Cholangitis Following Kasai Portoenterostomy

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Keywords

Anti-reflux valve
Ascending cholangitis
Biliary Atresia
Kasai portoenterostomy

Abbreviations

AC - Ascending cholangitis
ARV - Anti-reflux valve
BA - Biliary atresia
KPE - Kasai portoenterostomy

Abstract

Ascending cholangitis (AC) is the commonest and most serious complication following Kasai portoenterostomy (KPE) that significantly affects the long-term outcome of biliary atresia (BA). The role of anti-reflux valve (ARV) construction in preventing bile reflux into the Roux limb thereby reducing the risk of AC is debated in the literature. In this report we describe two infants who experienced AC within 1 month of KPE for BA. Blood cultures grew multi-drug-resistant organisms and imaging studies revealed bile lake formation. Both underwent the ARV construction to address persistent AC. One patient showed significant clinical improvement with no further episodes of AC over a year, while the other child rapidly deteriorated with the disease progressing to liver failure and death. While adjunctive therapies like ursodeoxycholic acid and prophylactic antibiotics are beneficial, ARV appears to be a promising option in reducing bile reflux and recurrent AC, thus prolonging the longevity of the native liver in BA.

INTRODUCTION

Ascending cholangitis (AC) is the commonest complication following Kasai portoenterostomy (KPE). It has profound impact on the prognosis of biliary atresia (BA).^(1,2,3) Recently, liver transplantation has emerged as the definitive treatment for BA. However, it is not available in several centers due to numerous challenges.⁽⁴⁾ This underscores the continuing importance of KPE in the era of transplantation and the need of developing strategies to mitigate complications such as AC and to preserve the native liver function for as long as possible.^(5,6)

Construction of an anti-reflux valve (ARV) is said to prevent the recurrence of AC by reducing bile reflux into the Roux limb. Thus, it has the potential to improve the long-term outcome, to enhance the quality of life and to delay the need for liver transplantation.^(7,8,9) Literature on the effectiveness of ARV is contradictory.⁽⁸⁻¹¹⁾ In this report, we share our experience with 2 infants who underwent ARV procedure as secondary intervention of multiple episodes of AC following KPE.

CASE REPORTS

Case 1

A term baby-boy, presented with prolonged conjugated hyper-bilirubinemia, pale stool and sonographic features suggestive of BA. On day-77 he underwent on-table cholangiography and KPE. He achieved jaundice clearance within 3 months of KPE (lowest serum total bilirubin 29.4 $\mu\text{mol/l}$). However, he developed 3 episodes of AC within a span of 4 months. Hence, ARV procedure was done 6 months after KPE. (Fig. 1) However, following the procedure, he rapidly deteriorated with onset of end-stage liver failure and succumbed to the disease a month later.

Case 2

A term baby-girl, presented with prolonged conjugated hyper-bilirubinemia, pale stools and ultrasonographic features of BA. On the day-85 of life intra-operative cholangiography confirmed type-3 BA and KPE was done. The lowest serum bilirubin level post-KPE was 118 $\mu\text{mol/l}$. She developed 3 episodes of AC within 4 months, for which ARV procedure was performed after 4 months of KPE. Following the ARV procedure, her clinical status improved significantly. There were no further episode of AC and her total bilirubin levels showed a consistent decline, reaching a nadir of 37.4 $\mu\text{mol/l}$ one year after KPE.

DISCUSSION

AC is the most prevalent and early complication of KPE.⁽¹⁾ Recurrent episodes of AC significantly increase the risk of liver fibrosis and cirrhosis, contributing to poor long-term outcome in BA.⁽²⁾ Early identification and timely surgical management of associated complications, such as the bile lakes, are crucial to preserve the native liver function and to delay the disease progression.⁽³⁾

Although KPE is not now considered curative of BA, it serves as a vital bridging procedure that delays the need for liver transplantation especially

in country with scarcity of donor liver.⁽⁷⁾ Consequently, optimizing KPE and additional interventions, such as the ARV procedure, becomes important to prolong the survival of the native liver and, to improve growth and nutrition while awaiting for eventual transplantation.

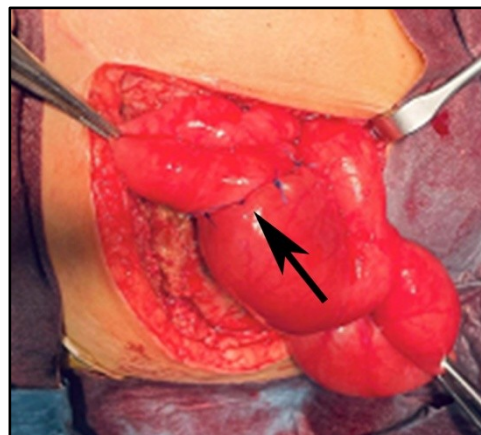


Fig 1. Construction of anti-reflux valve (arrow) in the Roux limb of portoenterostomy

ARV provides a relatively simple surgical solution for recurrent AC. Various types of artificial ARV include mucosal, conical, telescoping intestinal, and muscular valves.⁽⁸⁾ In our patients we employed conical valve technique. A one-way valve was constructed by intussuscepting a 3 cm segment of the Roux limb cranio-caudally near the previous jejunio-jejunostomy to prevent refluxing of the small bowel contents into the Roux limb. Non-absorbable monofilament sutures were used to secure the valve and its patency was tested. Case-1 highlights the severity of advanced liver disease, where even the ARV procedure failed to halt the onset of liver failure. Conversely, the successful clinical course of Case-2 underscores the potential benefits of ARV in mitigating recurrent AC and maintaining stable liver function.

Studies on the effectiveness of ARV are scanty reporting variable outcomes.⁽⁸⁻¹¹⁾ Nakajo performed ARV procedure on 23 patients, including those without AC.⁽⁸⁾ Among those with recurrent AC, 3

had complete resolution, 2 had reduced episodes and 1 died from persistent AC. Similarly, Saeki who studied ARV 20 infants, reported 1 death due to bile excretion failure and 1 case of recurrent AC but with fewer episodes.⁽⁹⁾ The long-term outcome of ARV in human infants is unknown. However, Nakajo suggested that atrophy or disappearance of the valve is unlikely due to its multilayered structure.⁽⁸⁾ In a postmortem examination Saeki reported of an ARV that maintained its structure and anti-reflux function for at least 2 years and 3 months.⁽⁹⁾

CONCLUSION

ARV appears to be a promising surgical option in the management of recurrent AC in BA. Further studies are warranted to evaluate the durability of the constructed ARV.

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