

## Introduction

- Congenital biliary atresia is a leading cause of neonatal cholestasis
- The Kasai is a surgical procedure (hepatportoenterostomy) involving radical excision of the biliary ducts and helps reestablish bile flow and attempts to prevent biliary cirrhosis.
- **The 20 year survival rate with native liver is less than 45%.**
- We present a unique case of a 31 year old male with biliary atresia who underwent the Kasai Procedure and has still not required a liver transplant despite his age.

## Case Description

- This is a 31 year old male with a known history of having biliary atresia status post Kasai procedure performed at (5 weeks old) presented for evaluation of abdominal pain.
- On the physical exam, he had jaundice and abdominal distention.
- His labs were pertinent for a total bilirubin of 3.1, alkaline phosphatase 187, AST 50, ALT 84, platelets 88, INR of 1.07 and albumin 4.2.
- An MRCP was performed to rule out cholangitis which showed postoperative changes of Kasai procedure, minimal intrahepatic duct dilation with marked left lobe atrophy and sequelae of hypertension.

## Image

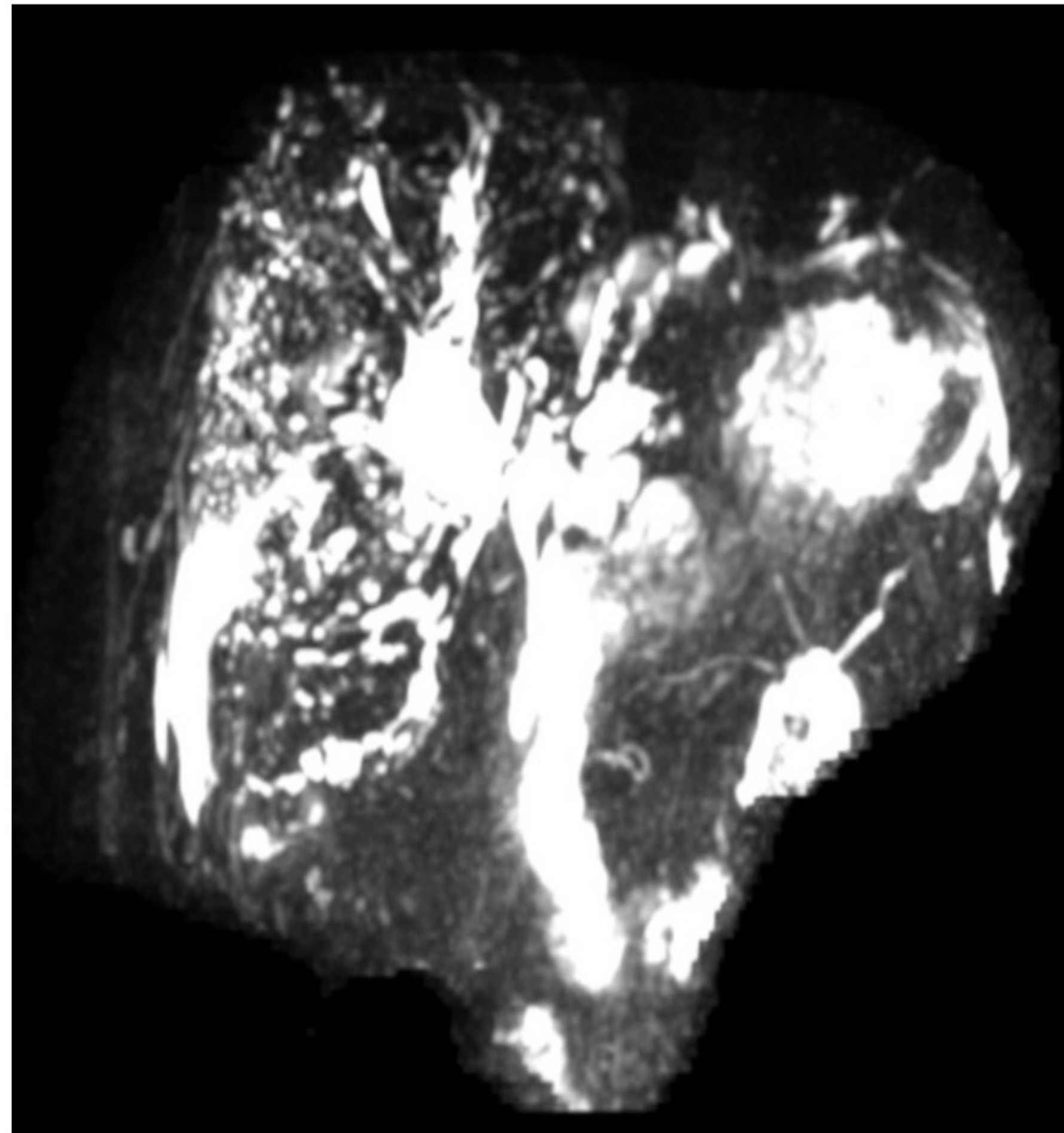


Figure: MRCP showing evidence of Kasai Procedure

## References

Hirzel, A. C., Madrazo, B., & Rojas, C. P. (2015, August 3). *Two rare cases of hepatocellular carcinoma after Kasai procedure for biliary atresia: A recommendation for close follow-up.* Case Reports in Pathology. Retrieved October 7, 2022, from <https://www.hindawi.com/journals/crpa/2015/982679/>

Khayat A, Alamri AM, Saadah OI. Outcomes of late Kasai portoenterostomy in biliary atresia: a single-center experience. *Journal of International Medical Research.* 2021;49(5). doi:[10.1177/03000605211012596](https://doi.org/10.1177/03000605211012596)

Toyosaka A, Okamoto E, Okasora T, Nose K, Tomimoto Y. Outcome of 21 patients with biliary atresia living more than 10 years. *J Pediatr Surg.* 1993;28:1498-501.

## Hospital Course

- treated with a course of intravenous antibiotic therapy and was diagnosed with cholangitis secondary to biliary stasis from his previous Kasai procedure
- followed by a Hepatologist who performed a fibroscan and diagnosed the patient with F4 fibrosis with Steatosis of S1 consistent with cirrhosis.
- MELD score was noted to be 10 and he was placed on ursodiol. A screening EGD was performed which showed grade 2 esophageal varices.
- The patient's plan is to eventually undergo a liver transplant in the near future.

## Discussion

- Common complications of patients with a prior Kasai include recurrent cholangitis, portal hypertension, and synthetic liver dysfunction which was seen in our patient.
- Prior case reports have shown that very few patients have survived longer than 20 years without a liver transplant.
- It is important for gastroenterologists to know the **long term side effects and complications that can be seen in these patients as they transition to adulthood.**
- To our knowledge and after an extensive literature review, **this is the longest survivor to date in the US**