

## A Case Report

# Successful Obstetric Outcome in a Woman with Prior Kasai Procedure for Biliary Atresia and Concurrent Alpha-1 Antitrypsin Deficiency: A Case Report

Ayodeji Oluwaseun Owolabi<sup>1</sup>, Mathew Koomson<sup>1</sup>

<sup>1</sup>Department of Obstetrics and Gynaecology. Royal Surrey County Hospital, Guildford, United Kingdom.

### Abstract

We present the case of a 33-year-old primigravida with a history of Kasai portoenterostomy at six weeks of age for congenital biliary atresia and co-existing alpha-1 antitrypsin deficiency. She received antenatal and intrapartum care in our maternity unit, in collaboration with a hepatologist at a tertiary facility. Her management involved a multidisciplinary team comprising maternal medicine and fetal medicine consultants, hepatologists, radiologists, and specialist midwives.

At booking (16 weeks), liver function tests were normal. She underwent four weekly growth scans and liver ultrasounds, with biochemical monitoring throughout her pregnancy. At 38 weeks, she presented with right upper quadrant pain over her surgical scar; clinical assessment and repeat liver tests were normal. Labour occurred spontaneously at 40 weeks and 2 days. She had a spontaneous vaginal delivery of a healthy female infant weighing 3.2 kg, with Apgar scores of 9 and 10 at one and five minutes. Both mother and baby remained well and were reviewed postnatally by the hepatology team.

This case is notable for demonstrating a successful term pregnancy and birth in a woman with rare dual hepatobiliary pathology (post-Kasai biliary atresia and alpha-1 antitrypsin deficiency) without hepatic decompensation. It highlights the importance of multidisciplinary coordination, close biochemical and fetal surveillance, and individualised antenatal planning in optimising maternal and neonatal outcomes. Informed consent for publication was obtained.

**Keywords:** Biliary atresia, Kasai procedure, alpha-1 antitrypsin deficiency, pregnancy.

**\*Correspondence:** Ayodeji Oluwaseun Owolabi, ([Owolabi.deji4@gmail.com](mailto:Owolabi.deji4@gmail.com)), Department of Obstetrics and Gynaecology. Royal Surrey County Hospital, Guildford, United Kingdom.

**How to Cite:** Owolabi AO, Koomson M. Successful Obstetric Outcome in a Woman with Prior Kasai Procedure for Biliary Atresia and Concurrent Alpha-1 Antitrypsin Deficiency: A Case Report. Niger Med J 2025; 66 (5): 2466-2469 <https://doi.org/10.71480/nmj.v66i6.1096>

Quick Response Code:



## Introduction

Biliary atresia is a rare neonatal condition characterised by progressive obliteration of the bile ducts. This often requires a Kasai portoenterostomy to restore bile flow.<sup>1</sup> Long-term survivors may face risks of liver dysfunction, with pregnancy potentially exacerbating hepatic stress. Alpha-1 antitrypsin deficiency further complicates management due to its association with liver and lung disease.<sup>2</sup> There is no published literature documenting pregnancy in women with both a history of Kasai surgery and co-existing alpha-1 antitrypsin deficiency. This report discusses antenatal care, delivery, and postpartum outcomes in a woman with both conditions.

## Case Presentation

A 33-year-old woman in her first pregnancy following spontaneous conception, presented for antenatal care at 16 completed weeks of gestation. Her medical history included congenital biliary atresia, for which she underwent a Kasai portoenterostomy at 6 weeks of age.

She also had alpha-1 antitrypsin deficiency. She was under annual outpatient follow-up and ultrasound scans by the Hepatologists at the tertiary hospital where the surgery was performed. Her most recent follow-up scan showed no active biliary atresia or hepatic dysfunction. She had been counselled about the possibility of liver transplantation if ageing or pregnancy aggravated her condition. Her husband tested negative for the carrier status of alpha-1 antitrypsin deficiency during pregnancy.

Plans were made for fetal medicine scans, early blood glucose monitoring (instead of an oral glucose tolerance test), and routine blood work, including a liver function test. The hepatologist at the tertiary centre where she received care evaluated her and found no immediate concerns. Four weekly liver function tests were recommended. An anomaly scan performed by a fetal medicine consultant at 20 weeks revealed no anomalies and normal uterine doppler results.

She had serial growth scans at 28, 32 and 36 weeks due to increased risks of preeclampsia and fetal growth restriction associated with alpha-1 antitrypsin deficiency. All laboratory investigations, including liver function tests, full blood counts, blood glucose monitoring and urine cultures, were normal throughout her pregnancy.

She remained well until 38 weeks, when she presented with mild, continuous right upper quadrant abdominal pain that was dull. There were no signs of jaundice or preeclampsia. The results of her blood tests, including liver enzymes and amylase, were reassuring. Her condition was managed conservatively, and she was discharged home one day later.

She presented in spontaneous labour at 40 weeks and 2 days and was monitored with a cardiotocogram due to intrapartum vaginal bleeding. She progressed well and had a spontaneous vaginal delivery. The baby was born in good condition, and the estimated blood loss was 500 ml. Both mother and baby were discharged home the following day with a referral to her hepatologist for postnatal review.

**Informed Consent:** Informed consent was obtained from the patient for the publication of this case report. All personal identifiers have been removed to ensure patient anonymity.

## Discussion

This case showed the possibility of an uncomplicated pregnancy and delivery in a woman with prior Kasai surgery and co-existing alpha-1 antitrypsin deficiency. In the Kasai operation, the surgeon removes the non-functioning extrahepatic bile duct and gallbladder, then connects the liver hilum to a loop of the jejunum.

As many as 60% of patients after Kasai surgery experience progressive liver-related complications.<sup>2</sup> The native liver survival rate after the Kasai procedure is 35–47% at 10 years.<sup>3</sup> Monitoring the decline in biological parameters after puberty is crucial to identify patients at risk for sudden hepatic insufficiency due to pregnancy.<sup>1</sup> Those with poor hepatic reserve are advised to complete liver transplantation before pregnancy.<sup>3</sup>

Pregnancy in such patients poses risks of severe liver dysfunction, portal hypertension and obstetric complications such as miscarriages and abruptio placentae.<sup>1,4</sup> The patient was counselled appropriately on these risks before and during pregnancy.

Alpha-1 antitrypsin deficiency is an autosomal recessive inherited disease affecting the lungs and liver.<sup>5</sup> The presence of a coexisting alpha-1 antitrypsin deficiency on its own has been linked to liver disease, preeclampsia, obstetric cholestasis, fetal growth restriction and preterm birth.<sup>6–8</sup> It is for this reason that serial growth monitoring has been conducted.

Pregnant women with pre-existing liver conditions may have impaired glucose metabolism. Oral glucose tolerance tests are avoided due to the significant glucose load that can strain the liver.<sup>9</sup> Hence, early blood glucose monitoring was used in this patient.

Her spontaneous vaginal delivery demonstrates the possibility of normal obstetric outcomes in carefully managed cases. This success story was achieved through multidisciplinary input from fetal medicine, obstetric teams, and a tertiary-level hepatologist.<sup>10</sup> The present case contributes to the limited literature regarding the pregnancy outcome of patients who have survived biliary atresia and who also have alpha-1 antitrypsin deficiency.

### Conclusion

Pregnant women with a background of Kasai surgery and co-existing alpha-1 antitrypsin deficiency can have favourable obstetric outcomes. Normal liver function, multidisciplinary care, and monitoring are essential for this to happen. This report emphasises the need for coordinated care when treating complex hepatobiliary and genetic conditions during pregnancy.

**Conflicts of interest** -Nil

**Funding for the study**- Nil

### References

1. Kuroda T. Pregnancy and Delivery in Adult Survivors of Biliary Atresia. In: Nio M (eds). *Introduction to Biliary Atresia*. Singapore: Springer; 2021 [cited 2025 Apr 22]. p. 289–94. Available from: [https://doi.org/10.1007/978-981-16-2160-4\\_41](https://doi.org/10.1007/978-981-16-2160-4_41)
2. Bijl EJ, Bharwani KD, Houwen RHJ, de Man RA. The long-term outcome of the Kasai operation in patients with biliary atresia: a systematic review. *Neth J Med*. 2013;71(4):170–3.
3. Hukkinen M, Ruuska S, Pihlajoki M, Kyrölahti A, Pakarinen MP. Long-term outcomes of biliary atresia patients surviving with their native livers. *Best Pract Res Clin Gastroenterol*. 2022;56–57:101764.
4. Sadiq J, Lloyd C, Hodson J, Trapero Marugan M, Ferguson J, Sharif K, et al. Long-term clinical and socioeconomic outcomes of children with biliary atresia. *JGH Open*. 2023;7(12):841–7.

5. Mornex JF, Traclet J, Guillaud O, Dechomet M, Lombard C, Ruiz M, et al. Alpha-1-antitrypsin deficiency: An updated review. *La Presse Médicale*. 2023;52(3):104170.
6. Orimoloye HT, He D, Li T, Janzen C, Barjaktarevic I, Wang X, et al. Alpha-1 antitrypsin deficiency and pregnancy complications and birth outcomes: A population-based cohort study in Denmark. *PLOS ONE*. 2024;19(1):e0296434.
7. Kosinski P, Kedzia M, Mostowska A, Gutaj P, Lipa M, Wender-Ozegowska E, et al. Alpha-1 Antitrypsin Z Variant (AAT PI\*Z) as a Risk Factor for Intrahepatic Cholestasis of Pregnancy. *Front Genet*. 2021;12:720465.
8. Tissarinen P, Tiensuu H, Haapalainen AM, Ronkainen E, Laatio L, Vääräsmäki M, et al. Maternal serum alpha-1 antitrypsin levels in spontaneous preterm and term pregnancies. *Sci Rep*. 2024;14(1):10819.
9. Rahim MN, Pirani T, Williamson C, Heneghan MA. Management of pregnancy in women with cirrhosis. *United European Gastroenterol J*. 2021;9(1):110–9.
10. Brady CW. Liver Disease in Pregnancy: What's New. *Hepatol Commun*. 2020;4(2):145–56.