A Life-Saving Liver Transplant in Salvaging a Case of Complicated Biliary Atresia - A Case Report

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Abstract

Background: Biliary atresia involves the progressive obliteration of the hepatic bile duct. It is a common cause of surgical jaundice in neonates, and is one of the commonest indications for liver transplant in children. The treatment is multidisciplinary, involving the surgical re-establishment of bile drainage into the intestine through a portoenterostomy. In certain patients with hepatic complications, liver transplantation is the definitive modality of treatment, however this is not readily available in Nigeria. We aim to share our experience and the surgical outcome in the management of a patient with biliary atresia who had a successful liver transplant.

Case Summary: A 2-year-old female child, who presented at 7 months of age on account of persistent yellowness of the eyes since birth, with associated passage of pale stool. Initial care was at different peripheral facilities, where she had been on medical therapy, with no improvement in symptoms, accounting for a delayed presentation to our teaching Hospital.

She had an exploratory laparotomy, with intraoperative findings of a hard craggy cirrhotic liver and an atretic gall bladder. Liver biopsy done showed cholestatic liver with cirrhosis.

She was subsequently referred to a facility in India where she had a successful liver transplantation done. Procedure was well tolerated with significant improvement in clinical and biochemical parameters, from a pre-operative conjugated bilirubin level of 11.4mg/dl (0-0.4mg/dl) to a current post-operative level of 0.2mg/dl

Conclusion and Recommendations: Liver transplant remains an important modality in the treatment of biliary atresia, with good surgical outcome. The management of biliary atresia however pose a challenge in Nigeria due to delayed presentation, and lack of facility for liver transplant. Despite these challenges, through an international collaborative effort, we were able to save the life of the patient and re-unite her with her family.

Keywords: Biliary atresia, Jaundice, Paediatrics, Liver transplant, multidisciplinary

Background

The occurrence of surgical jaundice in children is often associated with delayed presentation especially in developing countries such as Nigeria,[1,2] with this delay in appropriate management and intervention leading to development of life-threatening complications.[2] Biliary atresia accounts for one of the most common causes of surgical jaundice in neonates with an incidence of 1 in 15,000 live births.[3,4]

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It involves the progressive obliteration of the hepatic bile duct, and if progression continues without intervention, hepatic damage is imminent[5,6], accounting for one of the most common indications for liver transplant in children.[5,6]

The treatment is multidisciplinary and involves the surgical re-establishment of bile drainage into the intestine through a portoenterostomy (the Kasai procedure).[3]However only approximately 1/3 of patients survive in the long term with this procedure, while another 1/3 show no significant improvement following this surgery and progress to liver failure and die usually before the age of two years. In about another 1/3 of patients, the bilirubin level returns to normal and they show relatively normal growth for an indefinite period of time, however portal hypertension still develops, eventually requiring

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liver transplantation in the future.[3,7] Liver transplant has shown to be associated with good survival outcome.[8,9] Many developing countries such as Nigeria however currently have no facilities where liver transplant surgery can be performed, hence patients have to be refereed to international facilities to have their surgeries done. [10,11]

This is in contrast to advanced and developed countries of the world, where there has been remarkable improvement in the prognosis and surgical outcome of biliary atresia due to early presentation and intervention, as well as developed health infrastructures, where liver transplant surgeries are readily available.[12,13]

The average 10-year survival rate of children with biliary atresia who had primary portoenterostomies without liver transplant is estimated to be about 50%.[8] This is in comparison to patients who had liver transplant, having an overall 10 years survival rates of 82%, with an average corresponding overall graft survival rate of 71%.[14]

Studies have also shown that patients who had a primary liver transplant have superior long-term survival outcome compared with patients who had initial portoenterostomies and then subsequently had liver transplantation, showing the significant of liver transplant in the management of biliary atresia.[15]

The management of biliary atresia continues to pose a challenge in Nigeria due to late presentation, late referral, as well as inadequate infrastructure for liver transplant surgery.[1,2] To ensure the appropriate and successful management of biliary atresia, there is need for early recognition and intervention and an integrated multidisciplinary healthcare team approach in order to achieve optimal outcome.[16,17] It is therefore important to have a well-coordinated program between the paediatricians, the paediatric surgeons, the anesthetists and other healthcare stakeholders through a multidisciplinary, as well as international collaboration in the management biliary atresia, especially in developing countries such as Nigeria.

The objective of this article is to share our experience and the surgical outcome of a patient who had successful liver transplantation through a multidisciplinary and international collaborative effort, while highlighting the importance of liver transplant in the management of biliary atresia.

Case Summary

A 7-month-old female child, who presented to our facility at the University of Ilorin Teaching Hospital, Nigeria in the year 2022, on account of yellowness of eyes since birth, with asociated of passage of yellow-pale stool. Initial care had been at different peripheral facilities, where she had been on phototherapy, with no improvement in symptoms, accounting for a delayed presentation to our facility.

There was history of maternal fever in the late trimester, with history of resultant premature rupture of membrane. Child was delivered preterm via caesarean section at 33 weeks of gestational age with birthweight of 1.8kg. She is the second of a twin gestation, with no similar symptoms in her other twin sibling. She was fully immunized for age, with no delay in developmental milestones, and no other known co-morbity.

On examination; she was active, icteric, pale, afebrile, not dehydrated, with an appropriate weight of 6.8kg. Her abdomen was distended, but not tender, with the Presence of an umbilical and right groin hernia. Her liver was enlarged, 6cm below the right coastal margin, non-tender, with her diaper stained with pale stool.

She had a deranged liver function test, showing elevated conjugated bilirubin level, with a total bilirubin of 15.31 mg/dl (0.2-1), conjugated bilirubin of 11.4 mg/dl (0-0.4). ALT;175 IU/L (7-45), AST;185.86 IU/L (8-50), GGT; 149.26 IU/L (5-78), ALP; 971.3 IU/L

Full blood count result showed WBC of 13.35 * 10^{3}/mm^{3} with a PCV of 31.5%. Results of viral markers were negative. Renal function test and clotting profile were essentially normal. Abdominopelvic ultrasound scan done showed features suggestive of biliary atresia

We made an assessment of obstructed jaundice secondary to suspected biliary atresia, and the plan of management involved adequate counselling of the parents on the diagnosis and options of management. She subsequently had an exploratory laparotomy at 8 months of age with intra operative findings of a hard craggy cirrhotic liver and an atretic gall bladder. She sustained a rent in the supra hepatic IVC, about 1cm in length during liver mobilization, which was promptly repaired by the cardiothoracic team. She also had repair of an associated right inguinal and umbilical hernia. Liver biopsy done showed cholestatic liver with cirrhosis.

The immediate postoperative period was not adversely eventful. Postoperative serum electrolytes, urea, and creatinine were essentially normal; however, the full blood count revealed leucocytosis, with a white blood cell count of 19,000/mm^{3} and a packed cell volume (PCV) of 28%. On this account, she received a blood transfusion, with a post-transfusion PCV of 32%.

In the 12th day post operative period, she was noted to have spiking temperature, with persistent elevated WBC and progressive ascites, her weight at this time had increased to I9.6kg from initial 6.8kg. Samples for blood and urine culture, as well as malaria parasite teat were taken, and she was commenced on tab furosemide and spironolactone with significant improvement in ascites, however with worsening liver function.

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Her parents were adequately counseled on the diagnosis of liver cirrhosis and the need for liver transplant to save the life of the child, to which they consented to, and within one month, we were able to collaborate with an international facility in India; Amrita Institute of Medical Sciences and research center (AIMS), where she was referred to and eventually had a Live-donor liver transplant surgery done at the age of 8 months 29 days. The procedure was well tolerated and she was subsequently discharge on follow up. Her current liver function test two years post liver transplant shows a total bilirubin of 0.6 mg/dl (0.2-1), and a conjugated bilirubin of 0.2 mg/dl. (0-0.4), compared to a pre-operative conjugated bilirubin level of 11.4mg/dl (0-0.4mg/dl), as shown in figure 1.

All travelling and surgical expenses were made out-of-pocket by the parents, with an estimated surgical cost of two lakh sixty-three thousand five hundred and thirty-nine rupees (263,539Rs) = 4,561,195 naira.

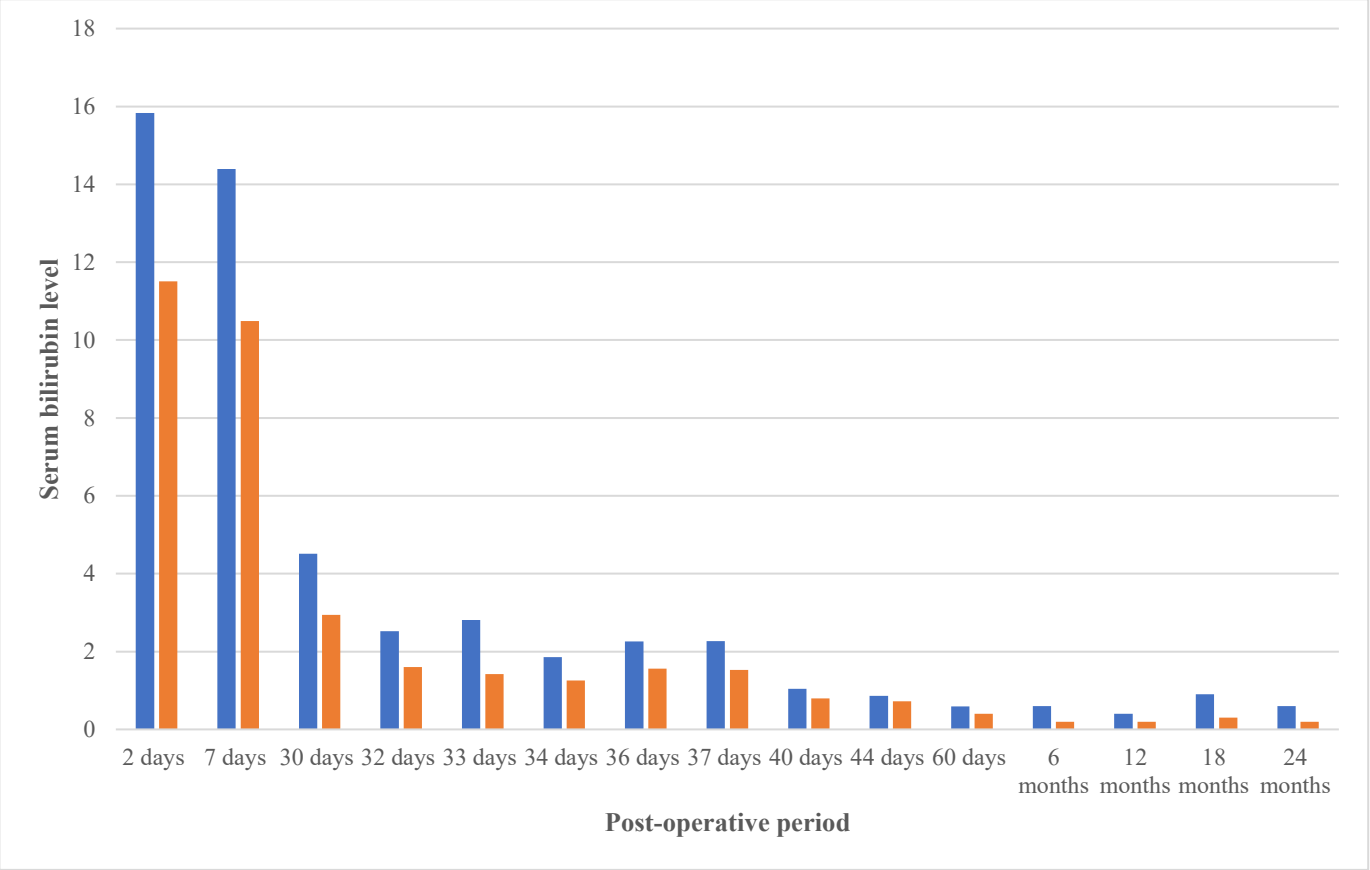

Figure 1; Total Bilirubin (mg/dl) and Direct Bilirubin (mg/dl) levels post liver transplant

Discussion

Studies have shown that over 70% of patients with biliary atresia present late, usually after the age of 3 months with associated complications, especially in developing parts of the world.[1,5] This is similar to our patient who had a delayed presentation, at 6 months of age. This was due to late referral, after being managed medically at different peripheral facilities, highlighting the need to create awareness and a multidisciplinary collaboration with other cadres of healthcare workers on the need for early diagnosis and prompt referral, given that surgical jaundice especially in the pediatric age group can be misdiagnosed and mismanaged as a medical pathology, thereby leading to delayed intervention and development of complications.[2]

This emphasizes the need to put together an inter-collaborative network between the pediatricians, the pediatric surgeons, and other healthcare stake holders in order to ensure prompt identification, referral and management of biliary atresia. If our patient had presented earlier, she might have benefitted from a portoenterostomy procedure, and may not have required the need for a liver transplant. However, it's important to also bring to light the fact that studies have shown that only about 30% of patient have a good long-term survival outcome with the use of portoenterostomies, with a higher chance of success in patients who have their interventions done within the first 2 months of life, before developing hepatic complications.[3,7]

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The survival rate with portoenterostomy is quite low, and given that the possibility of survival is not guaranteed, it is important to consider a more definitive option of management such as liver transplant. Patients who have liver transplant have been found to have an overall 10 years survival rates of 82%, and a corresponding overall graft survival rate of 71%,[14] hence patients have a better chance of survival with liver transplant compared to a portoenterostomy. Our patient is currently over two years post liver transplant, and she is alive and well, with significant clinical and biochemical improvement. In fact, she had started making remarkable and progressive decline in serum bilirubin level from the 30th day post-operative period as seen in figure 1. This is unlike in portoenterostomies where clinical and biochemical improvement may not be evident up until the 6th month post operative period.[15] This further agrees with the importance and supremacy of liver transplants as a modality of treatment in the management of biliary atresia over the use of portoenterostomies. Studies have also shown that patients who had a primary liver transplant have superior long-term survival outcome compared with patients who had initial portoenterostomies and subsequent liver transplantation.[15] It is therefore recommended that liver transplant should be seen as the primary modality of treatment for biliary atresia, with early diagnosis and prompt intervention provided.

However, many developing countries of the world such as Nigeria, are currently unable to perform liver transplant surgeries, due to the lack of adequate infrastructure, and therefore the need to refer patients to international facilities to have it done. In order to save the life of our patient, we had to refer her to an international facility in India, where she had her liver transplant surgery done. If these services were available in Nigeria, we would be able to provide prompt and cost-effective surgical intervention to our patients, and would not need to refer patients outside the country to have these surgeries done.

Conclusion

Liver transplant remains an important modality in the treatment for biliary atresia, with good surgical outcome. The management of biliary atresia continues to pose a challenge in Nigeria due to delayed presentation, and lack of facilities for liver transplant. Despite these challenges, through a collaborative effort, we were able to save the life of our patient and re-unite her with her family. In order to improve the surgical outcomes for children with biliary atresia, there is need for a multidisciplinary approach, as well international collaboration, while we continue to develop our nation's healthcare system.

References

1. Olusanya BO, Osibanjo FB, Mabogunje CA, Slusher TM, Olowe SA. The burden and management of neonatal jaundice in Nigeria: A scoping review of the literature. Niger J Clin Pract. 2016 Jan-Feb;19(1):1-17. doi: 10.4103/1119-3077.173703. PMID: 26755212.
2. Mshelbwala PM, Sabiu L, Lukong CS, Ameh EA. Management of biliary atresia in Nigeria: the ongoing challenge. Ann Trop Paediatr. 2007 Mar;27(1):69-73. doi: 10.1179/146532807X170538. PMID: 17469735
3. Arensman, Robert M. Paediatric Surgery. Landes Bioscience, 2009(2) :302-305
4. Kelly DA, Davenport M. Current management of biliary atresia. Arch Dis Child. 2007 Dec;92(12):1132-5. doi: 10.1136/adc.2006.101451. Epub 2007 Sep 18. PMID: 17878208; PMCID: PMC2066090
5. Okoro PE, Igwe P, Opara PI. Pattern and survival of biliary atresia patients; experience in southern Nigeria. Niger J Surg. 2013 Jan;19(1):4-6. doi: 10.4103/1117-6806.111495. PMID: 24027409; PMCID: PMC3762033.
6. Feldman AG, Mack CL. Biliary Atresia: Clinical Lessons Learned. J Pediatr Gastroenterol Nutr. 2015 Aug;61(2):167-75. doi: 10.1097/MPG.0000000000000755. PMID: 25658057.
7. Bijl EJ, Bharwani KD, Houwen RH, de Man RA. The long-term outcome of the Kasai operation in patients with biliary atresia: a systematic review. Neth J Med. 2013 May;71(4):170-3. PMID: 23723110
8. Fouquet V, Alves A, Branchereau S, Grabar S, Debray D, Jacquemin E, et al. Long-term outcome of pediatric liver transplantation for biliary atresia: a 10-year follow-up in a single center. Liver Transpl. 2005 Feb;11(2):152-60. doi: 10.1002/lt.20358. PMID: 15666395.
9. Superina R. Biliary atresia and liver transplantation: results and thoughts for primary liver transplantation in select patients. Pediatr Surg Int. 2017 Dec;33(12):1297-1304. doi: 10.1007/s00383-017-4174-4. Epub 2017 Oct 13. PMID: 29030698.
10. Garzali IU, Sheshe AA. Liver transplantation in Nigeria: The road so far. Japanese J Gastroenterol Res. 2022; 2(10):1095

https://doi.org/10.64061/tjhs.v33i2.6

11. Awuah WA, Ng JC, Bulut HI, Nazir A, Tenkorang PO, Yarlagadda R, Mustapha MJ, Abdul-Rahman T, Akilimali A, Oti VN, Aji N. The unmet need of organ transplantation in Africa. Int J Surg. 2023 Mar 1;109(3):519-520. doi: 10.1097/JS9.0000000000000025. PMID: 36927835; PMCID: PMC10389210.

12. Sadiq J, Lloyd C, Hodson J, Trapero Marugan M, Ferguson J, Sharif K, Mirza DF, Hirschfield G, Kelly D. Long-term clinical and socioeconomic outcomes of children with biliary atresia. JGH Open. 2023 Nov 10;7(12):841-847. doi: 10.1002/jgh3.12980. PMID: 38162865; PMCID: PMC10757476.

13. Holdar S, Alsaleem B, Asery A, Al-Hussaini A. Outcome of biliary atresia among Saudi children: A tertiary care center experience. Saudi J Gastroenterol. 2019 May-Jun;25(3):176-180. doi: 10.4103/sjg.SJG_306_18. PMID: 30479320; PMCID: PMC6526739.

14. Maria Hukkinen, Satu Ruuska, Marjut Pihlajoki, Antti Kyrönlahti, Mikko P. Pakarinen. Long-term outcomes of biliary atresia patients surviving with their native livers,Best Practice & Research Clinical Gastroenterology, Volumes 56–57,2022,101764,ISSN 1521-6918,https://doi.org/10.1016/j.bpg.2021.101764.

15. LeeVan E, Matsuoka L, Cao S, Groshen S, Alexopoulos S. Biliary-Enteric Drainage vs Primary Liver Transplant as Initial Treatment for Children With Biliary Atresia. *JAMA Surg.* 2019;154(1):26–32. doi:10.1001/jamasurg.2018.3180

16. Elebute OA, Timo MT, Alakaloko FM, Seyi-Olajide JO, Bode CO, Ademuyiwa AO. Biliary Atresia Splenic Malformation: A Case Report from A Tertiary Centre in West Africa and the Lessons Learnt. J West Afr Coll Surg. 2023 Jan-Mar;13(1):106-110. doi: 10.4103/jwas.jwas_195_22. Epub 2023 Jan 18. PMID: 36923807; PMCID: PMC10010588.

17. Mathur P, Gupta R, Soni V, Ahmed R, Goyal RB. Biliary atresia associated with polysplenia syndrome, dextrocardia, situs inversus totalis and malrotation of intestines. J Neonatal Surg. 2014 Jan 1;3(1):9. PMID: 26023480; PMCID: PMC4420428.

https://doi.org/10.64061/tjhs.v33i2.6