Nutritional Status of Children with Biliary Atresia and the Role of Portoenterostomy to Improve Nutrition

Shailesh Solanki, Veereshwar Bhatnagar, Sandeep Agarwala, Rakesh Lodha1, Nandita Gupta2, Manoj Kumar Singh3

Background: Nutritional depletion and growth stunting are present in patients with biliary atresia; “normal” nutrient and vitamin supplementation fail to correct these deficiencies. Children with this condition form the largest group for possible liver transplantation in the future; hence, stress should be laid on close attention to their nutrition.

Methods: Twenty-five patients with biliary atresia as cases and 25 age-matched children as controls were enrolled in the study from November 2010 to June 2012. Preoperatively, patients underwent standard investigations and anthropometric measurement (weight, height, and head circumference) assessment. Nutritional status (assessed with standard growth chart) was compared with control population, and children were divided into poor nutritional status and good nutritional status. Kasai’s portoenterostomy was performed in all patients, and comparison was done between preoperative nutritional status with postoperative status of children and also between hepatic iminodiacetic acid (HIDA) scan-positive (patent biloenteric pathway) children with HIDA scan-negative children. Postoperatively, after 12 weeks, the same anthropometric measurements were taken again, growth velocity (GV) was assessed, and children were divided into poor, average, and good GV.

Results: Nutritional status of children with biliary atresia was significantly poor than that of control group. Postoperatively, children had better nutritional status than preoperative nutritional status, especially in HIDA scan-positive children. GV was also significantly better in those children in whom postoperative HIDA scan was positive.

Conclusion: Children with biliary atresia have poor nutritional status in comparison to normal population and require multifaceted approach to achieve adequate nutrition. Establishment of a patent biloenteric pathway in these children improves their nutritional status and GV.

Keywords: Anthropometric measurements, biliary atresia, growth assessment, nutritional status
infants are very vulnerable to the debilitating effects of severe, prolonged malabsorption and ultimately require liver transplantation to sustain life. Nutritional management of these infants is difficult and requires meticulous attention. Here, we present observations and assessments related to nutrition in children with biliary atresia and effect of surgical outcome over growth parameters and velocity.

**METHODS**

The study was conducted in the department of pediatric surgery, at tertiary care center, over 2 years from November 2010 to June 2012. It was an observational, prospective study and started after clearance from the institute’s ethical committee. It included all cases of biliary atresia who underwent Kasai’s portoenterostomy. Children were excluded who had major bowel resection or other metabolic diseases, those on oral medication containing iron and vitamins, and those who were lost to follow-up or died before 3 months postoperatively. Our control group comprised 25 children of the same age group, with no hepatobiliary or gastrointestinal disease, and who were admitted in pediatric surgery ward during the same time period. Children were included in the study only after obtaining informed consent from parents. Preoperatively, all patients underwent standard imaging investigations (ultrasonography and hepatic iminodiacetic acid [HIDA] scan) and routine blood investigations, including liver function tests. Anthropometric measurements were done; parameters included (a) weight (kilograms) and weight percentile; (b) height (centimeters) and height percentile; and (c) head circumference (HC) (centimeters) and HC percentile.

For anthropometric assessment, K. N. Agarwal standard anthropometric growth charts were used. It has separate growth charts for boys and girls. “Poor” nutritional status was labeled if child’s anthropometric measurement value was <3rd percentile and “good” nutritional status if >3rd percentile.

Kasai’s portoenterostomy was performed in all children. Postoperatively, after 12 weeks, HIDA scan was repeated and anthropometric measurements were reassessed. Growth velocity (GV) was also assessed; it is a measurement of growth in fixed time duration. GV can be measured for any growth parameter such as GV for weight or height or HC. GV assessment was done postoperatively by the WHO GV assessment charts of 3 monthly intervals. “Poor GV” was defined if it was <25th percentile, “average GV” if it was >25th percentile but <50th percentile, and “good GV” if it was >50th percentile.

The data analysis was done using Stata software version 9.0 (Texas, USA), and Mann–Whitney test, Pearson’s test, and Fisher’s exact test were used. P < 0.05 was considered statistically significant.

**RESULTS**

A total of 50 children were enrolled in the study: 25 in the case group and 25 in the control group. There was a male preponderance among the studied children. The patient group (mean age = 3.14 months, range 1–5 months) had 22 males (88%) and three females (12%). In the control group (mean age = 3.52 months, range 2–5 months), there were 18 males (72%) and 7 females (28%). Both groups were comparable (P = 0.157). Postoperatively, 21 children had HIDA scan positive and four children had HIDA scan negative for bilioenteric drainage.

**Weight and height**

As depicted in Table 1. Preoperatively, 12 (48%) children had poor nutritional status for weight and 10 (40%) for height, while control group had none of the children with poor weight or height, and the difference was statistically significant (P = 0.0001). Postoperative group had 3 (12%) children with poor weight and 2 (8%) with poor height, and it was significantly better than preoperative group (P ≤ 0.05). In the postoperative group, there was significant improvement in nutritional status for weight and height of patients in whom HIDA scan was positive (P ≤ 0.05), but there was no statistically significant difference in status of patients in whom HIDA scan was negative (P = 0.157 and P = 0.317, respectively, for weight and height).

**Head circumference**

As depicted in Table 1. Preoperatively, 10 (40%) children had poor nutritional status for HC, while in control group, all children had good nutritional status (P ≤ 0.05). In postoperative group, 4 (16%) children had poor HC and the difference was not significant (P = 0.083) compared to preoperative group. There was significant improvement in nutritional status of patients in whom HIDA scan was positive (P = 0.002), but there was no statistically significant difference in nutritional status of patients in whom HIDA scan was negative (P = 0.083).

**Growth velocity**

We calculated GV for weight, height, and HC separately as depicted in Table 2. In 21 HIDA scan-positive children, 19%, 14%, and 14% had poor GV; 24%, 14%, and 9.5% had average GV; and 57%, 72%, and 76.5% had good GV, respectively, for weight, height, and HC. In HIDA scan-negative children, all 4 (100%) had poor GV for weight and HC. Only one child had average GV for height, and the rest three had poor. The difference in GV
Nutritional status of children with biliary atresia

Clinical assessment

- **Height**: 10%
- **Resting energy expenditure**: 10%

Interestingly, at initial presentation, the average child with biliary atresia has subnormal growth parameters. Saron et al. evaluated the nutritional status of pediatric age patients with autoimmune hepatitis and biliary atresia related to serum levels of Vitamins A, D, and E and the disease severity. They concluded that the highest nutritional deficit was observed in patients with biliary atresia, mainly with cholestasis. Similar results were found by Yu et al. and Mansi et al.; they concluded that children with liver disease have poor nutritional status and more severe in cholestatic conditions. Shiga et al. studied 45 biliary atresia patients and showed that patients of biliary atresia suffer from chronic hepatic dysfunction. They measured height, weight, triceps skinfold, midarm circumference, and midarm muscle area (MAMA). They concluded that MAMA was very useful parameter to characterize low metabolic status. In the present study, 48% of children had poor nutritional status for weight, while 40% of children had poor nutritional status for height and HC. All of these were statistically significant (P ≤ 0.05), i.e., more children of biliary atresia had poor nutritional status in comparison to controls. In the postoperative period, children (21 children) in whom biloenteric pathway was patent (HIDA scan-positive) showed significant improvement in nutritional status compared to HIDA scan-negative children. These findings confirm that biliary atresia children are nutritionally depleted and growth retardation is present, especially when cholestasis persists and liver disease continues to progress. Thus, growth of a child depends on many factors including fulfillments of protein-energy requirement along with macro and micronutrient intake. Many nutrients such as fat and fat-soluble vitamins need presence of bile in the intestine for absorption, and these all are essential for proper growth. Early in the disease, the child maintains normal nutritional status; however, as the disease progresses, the poor nutritional status becomes more obvious. Considering the pathology biliary atresia, children should receive appropriate nutritional care. Maintenance of good nutritional status in this group of children may lead to early stabilization for surgery, less postoperative complications (such as wound dehiscence and ascites), and early recovery in the postoperative period.

GV of children was assessed in the present study, children in whom the biloenteric pathway was patent (HIDA scan-positive) showed better GV in comparison to HIDA scan-negative children, and this was present in all three parameters, i.e., weight, height, and HC.

### Table 1: Anthropometric measurements of children with biliary atresia

| Serial number | Parameters                      | Number of controls | Number of cases (preoperative) | Number of cases (postoperative) HIDA +ve | Number of cases (postoperative) HIDA −ve |
|---------------|---------------------------------|--------------------|--------------------------------|------------------------------------------|------------------------------------------|
|               | $<3^{rd}$ % | $>3^{rd}$ % | $<3^{rd}$ % | $>3^{rd}$ % | $<3^{rd}$ % | $>3^{rd}$ % | $<3^{rd}$ % | $>3^{rd}$ % |
| 1             | Weight             | Nil               | 25                      | 12                      | 13                      | 1                      | 20                     | 2                      |
| 2             | Height              | Nil               | 25                      | 10                      | 15                      | -                      | 21                     | 2                      |
| 3             | Head circumference  | Nil               | 25                      | 10                      | 15                      | -                      | 21                     | 3                      |

$<3^{rd}$ %: Less than 3rd percentile according to K. N. Agarwal growth chart, $>3^{rd}$ %: More than 3rd percentile according to K. N. Agarwal growth chart, HIDA: Hepatic iminodiacetic acid Scan, +ve: Bilioenteric drainage present, −ve: No biloenteric drainage

### Table 2: Growth velocity assessment during postoperative period in children with biliary atresia

| Parameters | HIDA +ve number of patients | HIDA −ve number of patients |
|------------|-----------------------------|-----------------------------|
|            | Poor | Average | Good | Poor | Average | Good |
| GvWt       | 4    | 5       | 12   | 4    | Nil     | Nil  |
| GvHt       | 3    | 3       | 15   | 3    | 1       | Nil  |
| GvHC       | 3    | 2       | 16   | 4    | Nil     | Nil  |

HIDA: Hepatic iminodiacetic acid scan, GvWt: Growth velocity for weight, GvHt: Growth velocity for height, GvHC: Growth velocity for head circumference, +ve: Bilioenteric drainage present, −ve: No biloenteric drainage

### Discussion

The pathogenesis of growth failure and malnutrition in biliary atresia is multifactorial, with major contributions from increased energy expenditure and malabsorption related to cholestasis. Resting energy expenditure may be increased by as much as 30% in children with biliary atresia coupled with potential fat malabsorption, which correlates with the degree of cholestasis.

Failure to thrive, typically defined as poor somatic growth, is common in biliary atresia. Growth failure with below-average height and weight (z scores) was reported in 40% of patients in a large cohort of children with biliary atresia. A comprehensive retrospective multicenter analysis of another such group of children confirmed the prevalence of this problem and correlated failure to thrive with poor outcome. Clinical assessment of nutritional status in children with biliary atresia is less straightforward than it seems. Diminishment of height and HC GV is a late manifestation of poor nutritional status. Long-term developmental outcome is likely to be adversely affected by malnutrition, especially if there is poor head growth. Interestingly, at initial presentation, the average child with biliary atresia has subnormal growth parameters. Saron et al. evaluated the nutritional status of pediatric age patients with autoimmune hepatitis and biliary atresia related to serum levels of Vitamins A, D, and E and the disease severity. They concluded that the highest nutritional deficit was observed in patients with biliary atresia, mainly with cholestasis. Similar results were found by Yu et al. and Mansi et al.; they concluded that children with liver disease have poor nutritional status and more severe in cholestatic conditions. Shiga et al. studied 45 biliary atresia patients and showed that patients of biliary atresia suffer from chronic hepatic dysfunction. They measured height, weight, triceps skinfold, midarm circumference, and midarm muscle area (MAMA). They concluded that MAMA was very useful parameter to characterize low metabolic status. In the present study, 48% of children had poor nutritional status for weight, while 40% of children had poor nutritional status for height and HC. All of these were statistically significant (P ≤ 0.05), i.e., more children of biliary atresia had poor nutritional status in comparison to controls. In the postoperative period, children (21 children) in whom biloenteric pathway was patent (HIDA scan-positive) showed significant improvement in nutritional status compared to HIDA scan-negative children. These findings confirm that biliary atresia children are nutritionally depleted and growth retardation is present, especially when cholestasis persists and liver disease continues to progress. Thus, growth of a child depends on many factors including fulfillments of protein-energy requirement along with macro and micronutrient intake. Many nutrients such as fat and fat-soluble vitamins need presence of bile in the intestine for absorption, and these all are essential for proper growth. Early in the disease, the child maintains normal nutritional status; however, as the disease progresses, the poor nutritional status becomes more obvious. Considering the pathology biliary atresia, children should receive appropriate nutritional care. Maintenance of good nutritional status in this group of children may lead to early stabilization for surgery, less postoperative complications (such as wound dehiscence and ascites), and early recovery in the postoperative period.

GV of children was assessed in the present study, children in whom the biloenteric pathway was patent (HIDA scan-positive) showed better GV in comparison to HIDA scan-negative children, and this was present in all three parameters, i.e., weight, height, and HC.
Biliary atresia puts children at tremendous risk for caloric deficits, leading to failure to thrive. Physician caring for infants awaiting liver transplantation can do much, not only to prolong survival but also to maintain satisfactory growth and development. The key consideration is to provide adequate nitrogen and nonnitrogen calories and liberally utilize modern methods of enteral and parenteral alimentation when necessary either during preoperative or postoperative period.

**Conclusion**

Children with biliary atresia have poor nutritional status in comparison to normal population and require multifaceted approach to achieve adequate nutrition. Establishment of a patent bilioenteric pathway in these children improves their nutritional status and GV.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Balistreri WF, Grand R, Hoofnagle JH, Suchy FJ, Ryckman FC, Perlmutter DH, et al. Biliary atresia: Current concepts and research directions. Summary of a symposium. Hepatology 1996;23:1682‑92.
2. Kaufman SS, Murray ND, Wood RP, Shaw BW Jr., Vanderhoof JA. Nutritional support for the infant with extrahepatic biliary atresia. J Pediatr 1987;110:679‑86.
3. Khadilkar VV, Khadilkar AV, Choudhury P, Agarwal KN, Ugrn D, Shah NK. IAP growth monitoring guidelines for children from birth to 18 years. Indian Pediatr 2007;44:187‑97.
4. World Health Organization Multicentre Growth Reference Study Group. WHO child growth standards based on length/height, weight and age. Acta Paediatr Suppl 2006;450:76‑85.
5. McDiarmid SV, Anand R, Lindblad AS, Principal Investigators and Institutions of the Studies of Pediatric Liver Transplantation (SPLIT) Research Group. Development of a pediatric end-stage liver disease score to predict poor outcome in children awaiting liver transplantation. Transplantation 2002;74:173‑81.
6. Pierro A, Koletzko B, Carnielli V, Superina RA, Roberts EA, Filler RM, et al. Resting energy expenditure is increased in infants and children with extrahepatic biliary atresia. J Pediatr Surg 1989;24:534‑8.
7. Kobayashi A, Ohbe Y, Yonekubo A. Fat absorption in patients with surgically repaired biliary atresia. Helv Paediatr Acta 1983;38:307‑14.
8. Utteron EC, Shepherd RW, Sokol RJ, Bucuvalas J, Magee JC, McDiarmid SV, et al. Biliary atresia: Clinical profiles, risk factors, and outcomes of 755 patients listed for liver transplantation. J Pediatr 2005;147:180‑5.
9. DeRusso PA, Ye W, Shepherd R, Haber BA, Shneider BL, Whittington PF, et al. Growth failure and outcomes in infants with biliary atresia: A report from the biliary atresia research consortium. Hepatology 2007;46:1632‑8.
10. Sokol RJ. Medical management of the infant or child with chronic liver disease. Semin Liver Dis 1987;7:155‑67.
11. Cohran VC, Heubi JE. Treatment of pediatric cholestatic liver disease. Curr Treat Options Gastroenterol 2003;6:403‑15.
12. Shiga C, Ohi R, Chiba T, Nio M, Endo N, Mito S, et al. Assessment of nutritional status of postoperative patients with biliary atresia. Tohoku J Exp Med 1997;181:217‑23.
13. Saron ML, Godoy HT, Hessel G. Nutritional status of patients with biliary atresia and autoimmune hepatitis related to serum levels of vitamins A, D and E. Arq Gastroenterol 2009;46:62‑8.
14. Yu R, Wang Y, Xiao Y, Mo L, Liu A, Li D, et al. Prevalence of malnutrition and risk of undernutrition in hospitalised children with liver disease. J Nutr Sci 2017;6:e55.
15. Mansi Y, Ghaffar SA, Sayed S, El-Karaktsy H. The effect of nutritional status on outcome of hospitalization in paediatric liver disease patients. J Clin Diagn Res 2016;10:SC01‑5.