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TOTAL ABSENCE OF THE SMALL-BOWEL IN A NEWBORN WITH DUODENAL OBSTRUCTION A CASE REPORT AND COMPLETE REVIEW OF SHORT BOWEL SYNDROME

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ABSTRACT

Total absence of small bowel is very rare situation. It usually accompanied with vascular accident or abdominal wall defect. The following case is presented to discuss the optimal care and best operative technique could be had in this situation.

Case: Two-day-old neonate, G/A of 36th weeks, whom was referred to our center because of bilious vomiting and double bubble sign in the X-rays. Patient with diagnosis of duodenal obstruction was resuscitated and was transferred to operating room for exploration. We found adhesion and a sac, which was containing two blind pouches. The proximal was third portion of duodenum and the distal was cecum and 3 cm of terminal ileum. To end the operation duodenocolic anastomosis was performed. Patient transferred to NICU, for parenteral nutrition and discussing the matter with parents. Despite good postoperative course, patient was discharged on parent's consent on third days and died in home because of neglect.

Discussion: Patients with Short bowel syndrome can survive if at least 15 cm of small bowel with intact ileocecal valve or 25 cm small bowel without ileocecal valve are existed. If the above criteria having not been met, it is better to make bowel continuity back, put the patient on parenteral nutrition, then discuss the options of future transplant and costs, complications of prolong parenteralnutrition. These give them the time to adapt and decide

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INTRODUCTION

Total absence of small bowel is a very rare entity. It usually accompanied with vascular accident or abdominal wall defect. Usually in exploration a sac remaining of absorbed bowel and sign of previous inflammation and adhesion are obvious. Few cases of complete absence of small bowel without the evidence of previous pathology has been reported. We usually face with extremely short bowel that deciding let the patient die or keeping them alive by parenteral nutrition is very hard to make.

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The ***short-bowel syndrome*** is a disorder clinically defined by malabsorption, diarrhea, steatorrhea, fluid and electrolyte disturbances, and malnutrition. By adulthood, the small intestine grows from approximately 250 to 750 cm therefore, the infant and the young child have a favorable long-term prognosis compared to an adult regarding potential intestinal growth after intestinal resection possibly due to much more release of intestinal growth agents such as GLP-2,IGF-1 etc. There is much more problem when no bowel existed but as mentioned above these latter one is very rare entity we call it here *Ultra-Short-Bowel-Syndrome (USBS*). In this issue we don't want to discuss any more about the classic small bowel syndrome, but we are going to present USBS case and our management strategies for patient problem.

Case presentation

We present two-day-old neonate, G/A of 36th weeks, birth weigh 1800 gr. whom was referred to our center because of bilious vomiting and upper abdominal distension following delivery.At first the patient re-hydrated and NG Tube inserted which relieved the distension. Patient Lab. data were as following Hb 19, ABG PH 7.49, HCO3 25. Patient underwent routine radio logic study including supine / upright abdominal x-rays which showed double bubble sign. Because of x-rays Diagnosis of duodenal obstruction was made and sequentially transferred to operating room for exploration. Patient under general anesthesia and routine preparation through a supra-umbilical transverse incision explored. In exploration, adhesion and a sac, which was containing two blind pouches of small bowel was found. The proximal was third portion of duodenum and the distal was cecum and 3 cm of terminal ileum. Both pouches were fused together in sac and before dissection could not be identified. After discussing the problem with his family about the chance of survival and options which may help him, to end the operation duodeno-colic anastomosis was performed with 5/0 PDS suture. Patient transferred to NICU, for par-enteral nutrition. Patient had good postoperative course with no complication. He only developed severe diarrhea for which rehydrated with R/L. After a few days the parent noticed the difficulty and the time they should spend to keep him alive so, they made up their mind to release their patient on own consent. We followed the patient who died on 7th day of discharge because of severe dehydration.

DISCUSSION

The infant Patients with Short bowel syndrome can survive if at least 15 cm of small bowel with intact ileocecal valve or 25 cm small bowel without ileocecal valve are existed. Loss of the ileocecal valve may have important effects in patients with ileal resection (Cummings, 1984). The adaptation process is essential to understanding the patho-physiology of short bowel syndrome. Twenty-four to 48 hours after bowel resection in experimental animals, the remaining small intestine undergoes an adaptive response characterized by epithelial hyperplasia (Fine, 1983; Goulet, 1991; Grant, 1996 and Grosfeld, 1986). The nutrient effect appears to be regulated by feeding-induced release of one or more trophic gastrointestinal hormones, a stimulus that is mediated primarily by fat content of food. Glucagon-like peptide-II (GLP-2) induces marked villous hyperplasia within four days of administration in an animal model and has been demonstrated to have some benefits in human clinical trials (Hwang, 1986). The effects of various nutrients on intestinal adaptation have been studied in animals and humans. Animal studies have demonstrated reduced intestinal permeability with par-enteral arginine and glutamine supplementation (Scolapio, 1997; Spagnuolo, 1996) glutamine provides modest benefit in body weight, fluid and electrolyte balance (Sial, 1994). Long-chain triglycerides appears to be more beneficial in promoting overall intestinal adaptation than supplementation with medium-chain triglycerides, even though medium chain triglycerides may be more easily absorbed (Hwang, 1986 and Thompson, 1993). The likelihood of successful transition to enteral feeding in short bowel syndrome depends upon several factors (Bloom, 1987; Buchman, 1997; Byrne, 1995): The length of remaining small bowel, the remaining segments of intestine continuity, Presence of the colon and an intact ileocecal valve, finally grade of Intestinal adaptation. Aforementioned factors are true for all kinds of short bowel syndrome induced by different mechanism but sometimes as we discussed in this case report we encounter with rare patients with ultra-short-small bowel segment in which you do not have wide range of maneuver to perform in order to elongate this diminutive bowel segment so we should consider all possible choices into effort but we insist on complete discussion of patient survivability and possible long-term complications of processes to parents in law.Then the second most important in these special group of patient is sepsis and dehydration prophylaxis; never make your final therapeutic decision solely based on patient's condition on presentation, first of all resuscitate patient and bring water and electrolyte in normal range then this is the fact that when we do not have any absorption area so we should start hyper alimentation as soon as possible in order to maintain patient stable but keep in mind that TPN is not the final goal but the tool in order to reach there besides long term hyperalimentation causes some serious problems for patients. in USBS we should consider multi-interventional challenge at least for research basis. So, we can administer bowel growth stimulating agents such as GLP-2, IGF-1, Glutamine oral simultaneously with operative management.

Our goals in operative management are bowel course continuity, for which after minimal amount of resection to reach anastomaseable margins. We should try to anastomose two ends as we did in this case but keep in mind that secure anastomosis in these severely immunocompromised patients does not need any more insist you can reach to latter goal by anastomosis in layers (instead of one layer with separate sutures) or securing anastomosis with bio-glue, try to elongate Bowel course by different procedures such as classic STEP or LILT or some investigational techniques such as skin or other tissue tubularization and possibly intestinalization (which is just our hypothesis) also try to make delay in diminutive small bowel course transit by making surgical ileocecal valve for example by folding mucosa and underlying soft tissue layers, finally consider small bowel transplant. In this context we should mention that selection of patients with ultra-short-bowel syndrome for small bowel transplant should be based upon special criteria and just after complete stabilization of patient in regard to sepsis, non-surgical methods and non-transplant surgical techniques .before patient undergo small bowel transplant we should completely assess and reinforce their nutritional status in order to decrease mortality and increasing success rate.

Important goals in this regard are

-1. Prevent or treat malnutrition and establish a positive nitrogen balance-
-2. Optimize nutritional status by achieving desirable body weight, muscle mass, and visceral protein stores
-3. Monitor and supplement vitamin and mineral levels to prevent deficiency.
-4. Avoid potentially toxic substances
-5. Improve nutritional status to counteract the catabolic effects of surgery and high-dose immunosuppression
-6. Limit the risk of complications, such as hypoglycemia and infection
-7. Promote growth and development.
-8. manage nutritional adverse effects of immunosuppressive therapy.
-9. Support wound healing
-10. Optimize the patient's quality of life.

Nutritional assessment should be started early and monitored regularly; it is based on complete medical history taking, physical examination for signs of nutrient deficiencies or toxicities, and biochemical measurements of nutritional status. Reassessment should be completed at least every 3-4 months. Kilocalorie requirements can be estimated by using standard nomograms, such as the Recommended Daily Allowance (RDA). Although these values apply to most children, the data may need to be adjusted if patients have severe malnutrition. Especially in USBS group. Growth charts are analyzed to determine the type and degree of malnutrition. Chronic malnutrition is manifested as stunting. This is best determined by performing serial determinations of the height-age index. (Tab- 1)

bowel transplant can be made isolate or in conjunction with liver transplant or in conjunction with liver and pancreas which we call it multivisceral transplant.In most SBS and certainly all USBS cases because of chronic TPN liver steatosis and possibly failure may be present so before performing isolate small-bowel transplant we should assess liver function and structural change adequately. Source of small bowel transplant are now heart-beating donors but with further evolution of non-hepatotoxic more potent immunosuppression drugs we should be hope full for possible porcine or etc. source small bowel transplant. Absolute contraindications to intestinal transplantation include congenital immunodeficiency syndromes because of the risk of unrestrained graft versus host disease (GVHD).

Tab. 1. Estimated Energy Requirements in Infants and Children

Age, y	Kilocalories, kcal/kg body weight
0-1	90-120
1-7	75-90
7-12	60-75
12-18	30-60

Tab. 2. Estimated Protein Requirements in Infants, Children

Age, mo.	Protein Requirement, g/kg
0-6	2.2
6-12	2

Tab. 3. Daily Fluid Requirements of Pediatric Patients

Group and Weight, kg	Fluid
Premature neonates, <2	150 mL/kg
Neonates and infants, 2-10	100 mL/kg for first 10 kg

Tab. 4. RDAs and Adequate Intakes for Fat-Soluble Vitamins

Category	Age or Time, y[†]	Vitamin A, mcg[‡]	Vitamin D, mcg[*§]	Vitamin E, mg[‖]	Vitamin K, mcg[*]
Infants	0.0-0.5	400[*]	5	4[*]	2
	0.5-1	500[*]	5	5[*]	2.5
Children	1-3	300	5	6	30
	4-8	400	5	7	55

Tab. 5. RDAs for Water-Soluble Vitamins

Category	Age or Time, y[†]	Vitamin C, mg	Thiamine, mg	Riboflavin, mg	Niacin, mg[‡]	Vitamin B-6	Folate, mcg	Vitamin B-12, mcg
Infants	0.0-0.5	40[*]	0.2[*]	0.3[*]	2[*]	0.1[*]	65[*]	0.4[*]
	0.5-1	50[*]	0.3[*]	0.4[*]	4[*]	0.3[*]	80[*]	0.5[*]
Children	1-3	15	0.5	0.5	6	0.5	150	0.9
	4-8	25	0.6	0.6	8	0.6	200	1.2

Tab. 6. RDAs and Adequate Intakes for Minerals

Category	Age or Time, y[†]	Calcium, mg[*]	Phosphorus, mg	Magnesium, mg	Iron, mg	Zinc, mg	Iodine, mcg	Selenium, mcg
Infants	0.0-0.5	210	100[*]	30[*]	0.27[*]	2[*]	110[*]	15[*]
	0.5-1.0	270	275[*]	75[*]	11[*]	3	130[*]	20[*]
Children	1-3	500	460	80	7	3	90	20
	4-8	800	500	130	10	5	90	30

Nitrogen balance studies, such as the 24-hour urinary urea nitrogen test, are the standard criteria for assessing protein needs. When these studies are not feasible, estimation based on the RDA for age can be used. These values serve as baselines and may need to be adjusted for the patient's state of malnutrition and/or physiologic stress (Tab- 2, 3, 4, 5, 6). When TPN support cannot be maintained because of complications such as advanced liver disease, loss of venous access, or central line sepsis (TPN complications), small-bowel transplantation becomes a therapeutic option. Small

Relative contraindications are evolving, but concern appears to be growing about transplantation from CMV- or EBV-positive donors to CMV- or EBV-negative recipients. These concerns are caused by the high morbidity and mortality rates associated with the development of CMV or lymphoproliferative disease in pediatric intestinal transplant recipients. No clear lower age limit for intestinal transplantation has been established for pediatric patients. Although critically ill patients should be excluded, a history of multiple abdominal surgeries is not a contraindication for transplantation. *Hakim* (1999) reports a

recommendation to retrieve a small-intestine graft from a stable cadaveric donor with a beating heart who is 20% smaller than the recipient. This recommendation is designed to ensure that the pediatric recipient (who is likely to have a contracted peritoneal cavity) can accommodate the graft. The shortage of potential donors because of size constraints is prompting development of novel harvesting and grafting techniques. Technical errors are more common in children than in adults the errors may cause graft loss. The errors include anastomotic leaks, hepatic artery thrombosis, and biliary anastomosis leaks in combined transplantations another option for possible more effective small bowel transplant are serial bowel anastomosis in antiperistatical fashion in order to increase small bowel transit time and let bowel to absorb most possible of contents also in investigational models we can use some electrical pacing in orderto slow down transit or reverse bowel movement direction.

Conclusion

Finally, here we can summarize our approach to: stabilization, hyperalimentation, non-surgical intervention, non – transplant trial surgical intervention, human and non-human kind small bowel transplantation. Here we should emphasize the importance of financial efforts at least by research programs to support these special infants.

Disclosure

The authors declare no conflicts of interest.

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