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Case Report

Multi-Step Treatment of Ultra-Short Bowel Syndrome with Unclear Outcome in a Newborn with Prenatal Known Gastroschisis. A Case Report

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Abstract

Presently, there is not much literature on therapy of severe gastroschisis with concomitant ultra-short bowel syndrome in newborn, even though the numbers of patients are rising. For an early starting treatment, there is even no data at all. We are presenting the case of a premature born child who had severe gastroschisis with a type 3b atresia of the small intestine and a resulting ultra-short bowel syndrome. It is usually recommended to wait some time with the therapy, but there is no concrete strategy or guideline in case waiting is not an option. For this reason, we now present our approach with an early serial transverse enteroplasty procedure, which entailed a positive result.

Keywords: *Premature Baby; Ultra-Short Bowel Syndrome; Gastroschisis; Serial Transverse Enteroplasty Procedure; Nutrition*

Introduction

Gastroschisis, along with omphalocele, is one of the two most common congenital abdominal wall defects [1] with a current incidence between 1.79 and 1.97 per 10000 births in Germany [2,3]. Nevertheless, early therapeutic approaches are hardly explored. In contrast to an omphalocele, in which the intact hernia contains amnion, peritoneum and portions of the intraperitoneal organs, the exuding abdominal organs in Gastroschisis are not covered by a membrane, which complicates the therapy in the further course. Various potential triggers for the development of a gastroschisis are described, but so far none of these hypotheses provide an adequate explanation for how it ultimately comes to a defect of the abdominal wall [4]. Children with gastroschisis are often born too young and too small, which makes therapy considerably more difficult, but advances in pediatric surgery and neonatal intensive care have helped to increase the overall survival rate of the affected children to over 90% [5,6]. The mortality rate of children with Gastroschisis, especially in complex cases, is high. Liver failure (60%) and sepsis (10 - 20%) are the most common complications. Moreover there is a significant coincidence between severe cases of gastroschisis and the occurrence of a short bowel syndrome [7-9]. Due to the trend in rising case numbers [10], the consequences of gastroschisis such as short- and ultra-short bowel syndromes have an impact not only on the children and their families affected, but also on the health care and cost recovery of hospitals [5]. So far there are rare experiences of treating gastroschisis in a premature baby with an additional Serial Transverse Enteroplasty procedure to treat ultra-short bowel syndrome and there is no such early attempt to be found in literature. Herein, we report a successful outcome of our therapeutic approach.

Case Presentation

We present the case of a male newborn, who was born in the 36th pregnancy week due to primary section because of a premature rupture of the membranes in known gastroschisis (Figure 1/Table 1A). At birth, the infant weighed 2500g at a height of 48 cm and had an

inconspicuous primary adaptation. Nevertheless, it was intubated early because of the gastroschisis and placed in the operating room in respiration. There, it presented itself rosy with APGAR 9/9/9, NapH 7.31, body temperature 36.8°C and BE -1.7 mmol/l. Apart from the gastroschisis there were no other visible malformations. Paraumbillically right a necrotic intestine of approximately 4 cm in length with small atretic intestinal buds and a mesenteric compound could be seen, fixed to the abdominal wall (Figure 2).

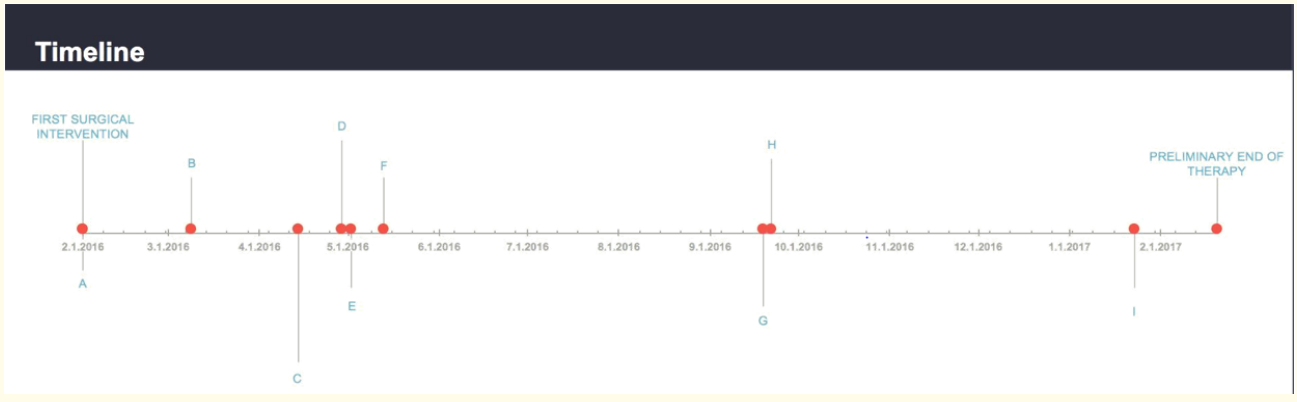

Figure 1: *Timeline.*

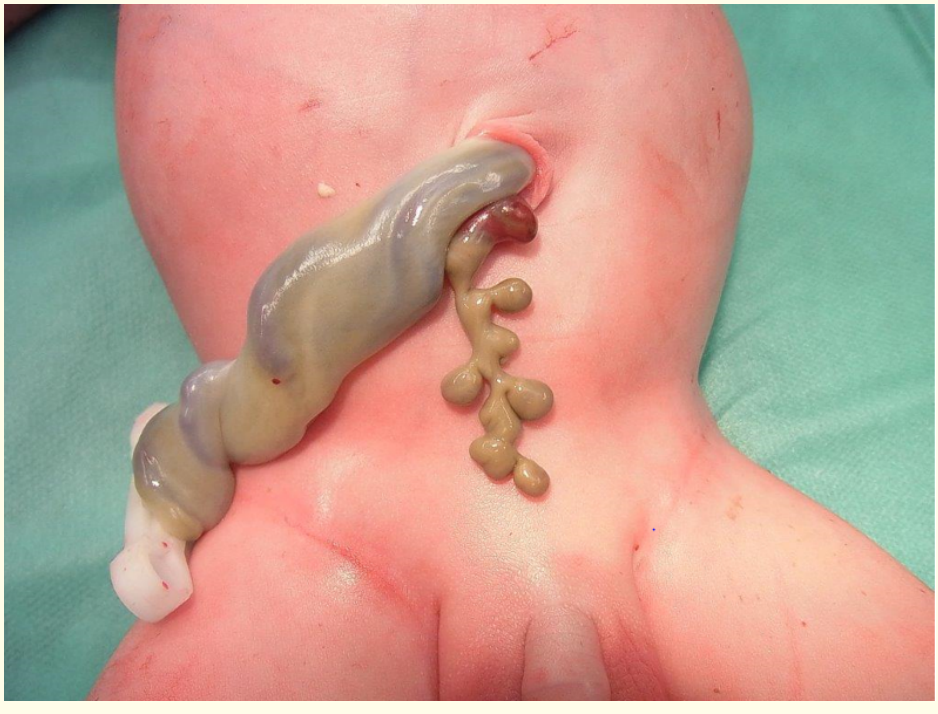

Figure 2: *Gastroschisis and visible small intestine buds at time immediately after birth.*

Initially we opted for a median laparotomy with the involvement of the former gastroschisis gap. Then, a cautious release of the mesenteric portion of the necrotic intestine with a multi-layered opening of the abdominal wall and exposure of the mesentery was done and a 3 - 4 cm wide loop of the small intestine (Figure 3) could be demonstrated and identified as type 3b atresia (Figure 4). The small intestine was spread out to the stomach, then the gastric secretions were aspirated. Since Treitz's ligament was not present, it was difficult to identify the blind-ending small intestinal piece as a proximal stump. Even the distal stump was difficult to find, but could then be represented as a thinly formed colon, which was not fixed retroperitoneally. Subsequently, a stripping of the clearly dilated proximal intestine portion and both bowel ends were discharged separately at the abdominal wall as a terminal anus praeter. The atretic part of the large intestine was discharged on the left side of the navel and, analogously, the portion of the small intestine paramedian on the right.

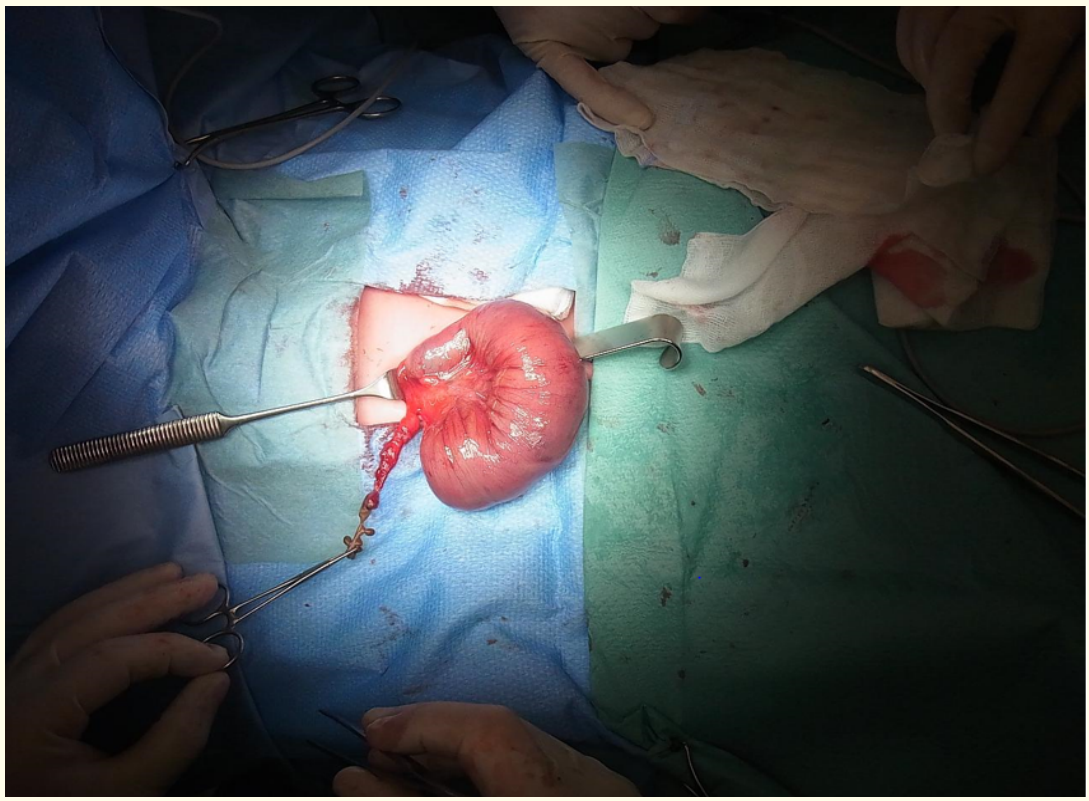

Figure 3: *Intraoperative view on type 3b atresia.*

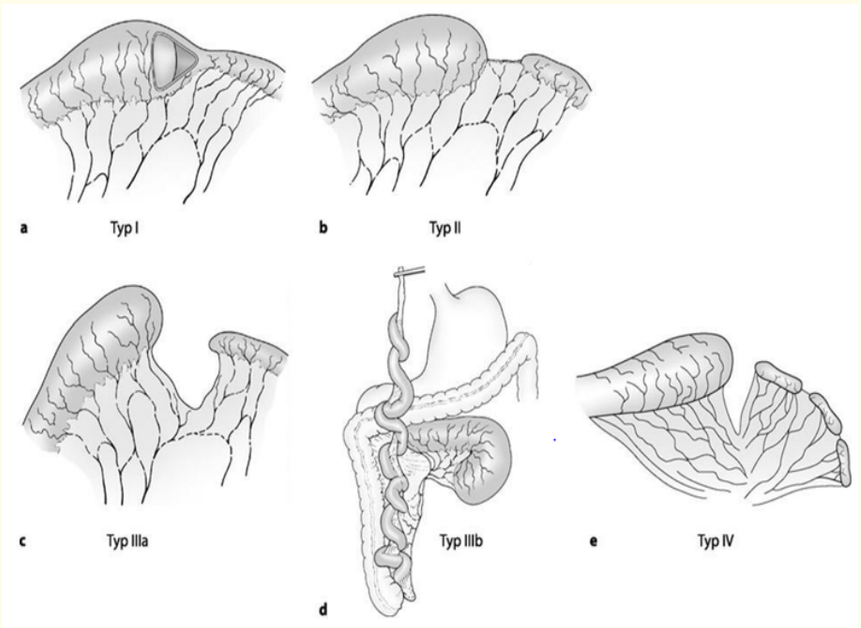

Figure 4: *Overview of small intestinal atresias; Schweinitz/ Ure. Lehrbuch „Viszerale und Allgemeine Chirurgie des Kindesalters", Springer-Verlag, 2009.*

Approximately one month after initial operative care (Timeline/Table 1B), the anus praeter was relocated with end-to-end anastomosis and elongation of the large intestine using the serial transverse enteroplasty procedure. At the beginning of the preparation of the small intestine a massively overgrown intestine could be found. The subsequent length measurement of the small intestine showed a length of 27cm antimesenterial, starting from the pylorus of the stomach with significant dilatation of the small intestine. Examination of the colon showed a loss of the colon in the area of the descending colon. The latter was dissected and sutured to the jejunum by end-to-end anastomosis after resection of the AP areas. After performing the STEP, the small intestine reached a length of 42 cm (Figure 5). Parallel to this, a Hickman catheter was installed via the jugular vein and a gastrotube was placed into the gastric antrum.

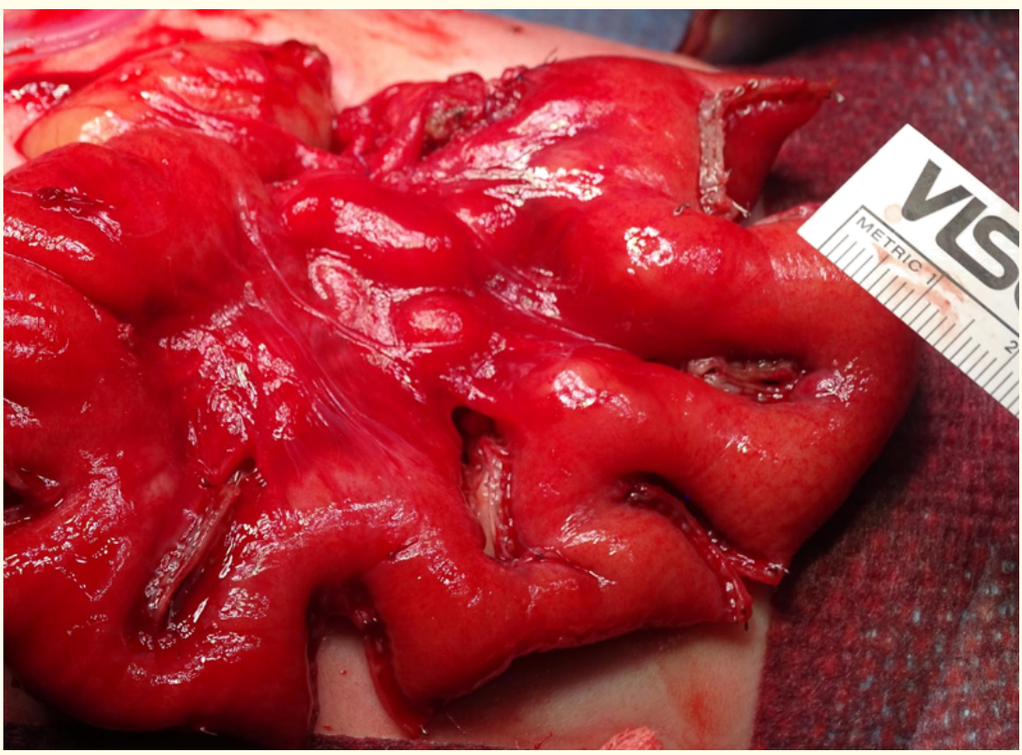

Figure 5: *Result of the first STEP.*

One month after the second surgical intervention (Timeline/Table 1C) a revision laparotomy was performed with a once again STEP procedure, revealing of small intestine adhesions and an exchange of the gastrotube. At this time the small intestine was clearly dilated, the wounds of the STEP procedure had healed and no classical change in the caliber could be seen. Only the caliber leap to the colon was still visible, which did not hinder the free passage of food. Another four forklifts were used, 3x jejunal and 1x at hilum (Figure 6). The result of both STEP can be seen in figure 7. The small intestine now came to a length of 60 cm.

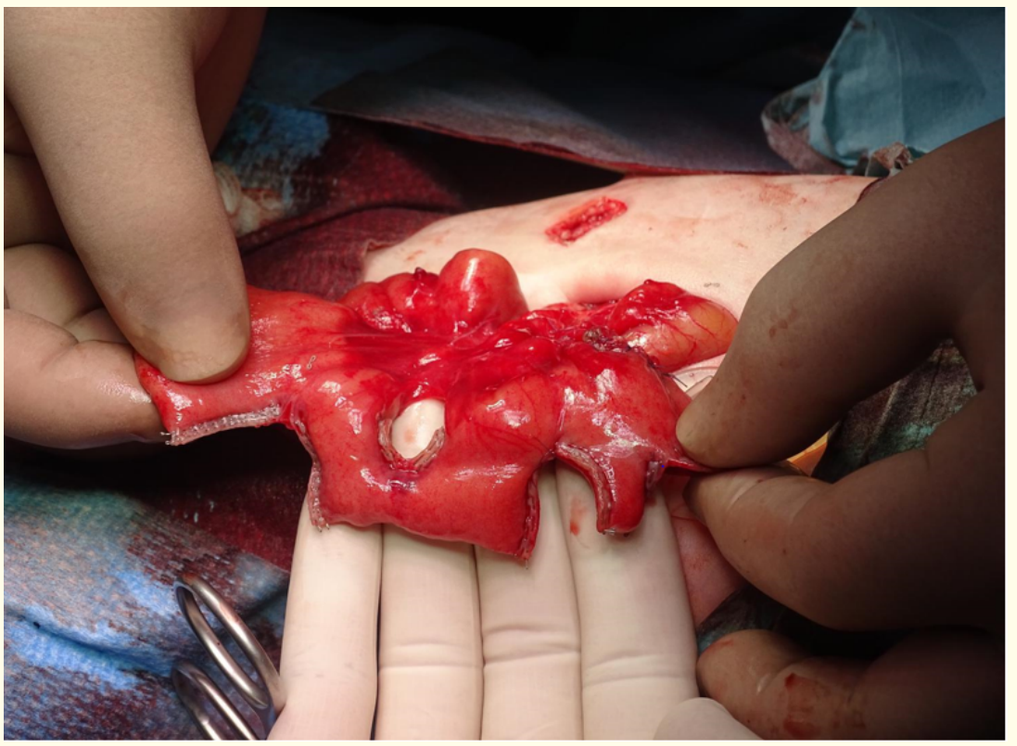

Figure 6: *Result of second STEP.*

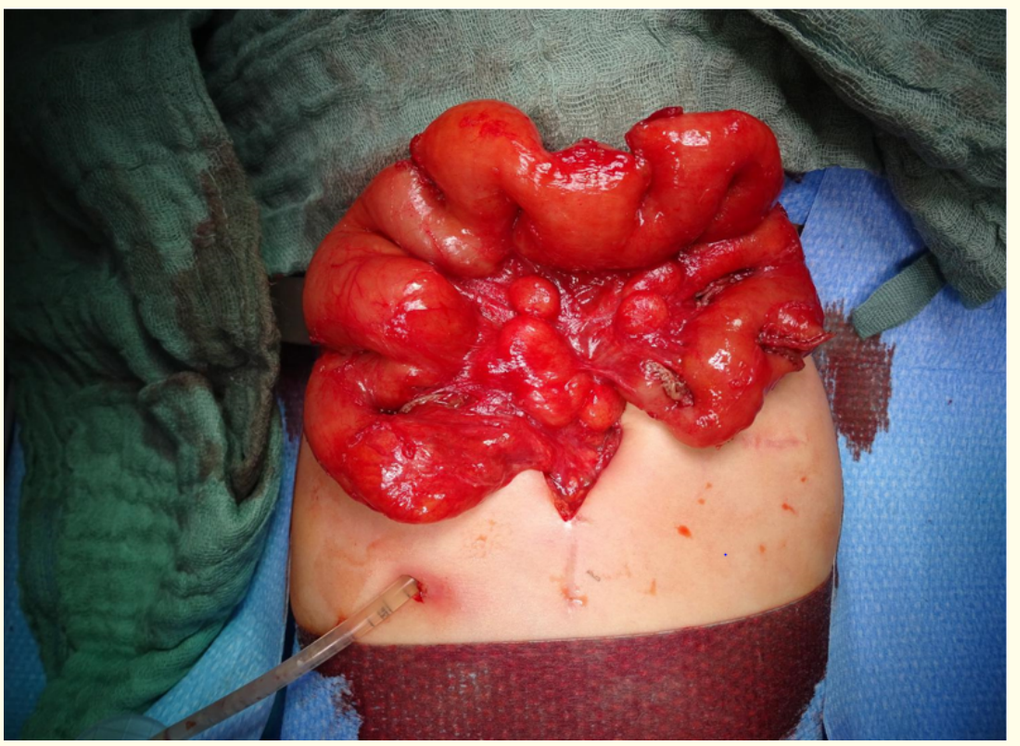

Figure 7: *Complete small intestine after both STEP.*

Following the second operation (Timeline/Table 1D, 1E), the child developed a catheter associated sepsis which we removed operatively. Despite negative evidence a prophylactic antibiotic therapy using cefuroxime, vancomycin and metronidazole was performed. After successful rehabilitation and therapy of sepsis, a new catheter was inserted three days later.

Ten days after we treated the sepsis (Timeline/Table 1F), the child developed a mechanic ileus. Since a clarifying presentation was not possible by laparoscopy, we decided to perform another median laparotomy. Preparatively the situation continued to be difficult, since new adhesions could be found after the STEP procedure. In addition, the small intestine convolute was found to be tilted by over 180° and an incomplete volvulus, with considerable bridging at the transition from small to large intestine, was present in the area of the anastomosis. This gluing was dissected and released, resulting in a transfer of small intestinal contents into the colon. Additionally, we ordered histologic examinations to exclude a nervous innervation disorder in this area. The old anastomosis persisted to prevent another loss of small intestine in the return procedure and was now reapplied 4cm distally as a double-lobular colon stoma. Finally, a change of the PEG probe and an anatomical occlusion of the abdomen took place.

Five months later (Timeline/Table 1G, 1H), the relocation of the double-lumen colonic stoma occurred, which led to a suture insufficiency with increased inflammatory parameters. There was an immediate antibiotic therapy with gentamycin/metronidazole and an operative care by revision of the AP three days later. Three months after this AP revision (Timeline/Table 1I), it was finally possible to relocate the double-lumen colonic stoma and produce an end-to-end anastomosis with jejuno-descendotomy.

A --> 02/2016	Laparotomy with resection of the atretic small bowel, installation of an anus praeter
B --> 03/2016	AP relocation with end-to-end-anastomosis; first STEP
C --> 04/2016	Laparotomy with second STEP
D/E --> 05/2016	Catheter removal and new installation because of catheter associated sepsis
F --> 05/2016	Laparotomy with detachment of adhesions, detorsion of small bowel volvulus and installation of a descendostoma
G/H --> 22.09.2016	Laparotomy with relocation of stoma/revision of AP after anastomotic leak
I --> 01/2017	Laparotomy with AP relocation

Table 1: *Time course of surgical interventions.*

The patient has since been clinically unremarkable and eating is unrestricted in choice and quantity. At the recent routine follow up at two years of age, a three-day lasting nutrional protocol revealed a sufficient intake of 1600 kcal/d (=178 kcal/kg/d). Intestinal adaptation is progressing as he has 3 - 5 times stool per day of soft to mushy consistency and volume and infusion time for parenteral nutrition was possible to be reduced over time. Under this regime both body weight and length is developing along the 3rd percentile. In detail, he is receiving 98 kcal/d via partly parenteral nutrition overnight, from 8:30 pm until 8:30 am (120 ml/kg BW), via the Hickman catheter (amino acids 2,5 g/kg/d, glucose 13,9 g/kg/d, fatty acids (SMOF) 3,3 g/kg/d). Laboratory testing of electrolytes, blood gas analysis, creatinine, BUN, liver enzymes, conjugated bilirubine and cystatine C were normal as well as the trace elements. The contrast intestinal passage performed at 25 months of age demonstrated a residual length of 55 cm small bowel and 19 cm of descendent colon and rectum without obstructions. The maximum width of small bowel lumen was 4 cm where the STEP procedure was performed.

Discussion

In order to understand how it came to our decision for an early surgical therapy of this patient with gastroschisis and ultra-short bowel syndrome, it is important to understand the problem of the present case. In our patient, there was a gastroschisis with concomitant type 3b intestinal atresia (Figure 6), which was expressed in a barely formed small intestine with a length of just 27 cm,

measured starting from the pylorus of the stomach. Basically, the gastroschisis is clinically differentiated into a simple and a complex course of the disease, which differs significantly in terms of further clinical care, postoperative complications, as well as the length of stay in the hospital and the mortality rate. The latter is 0% for simple cases and 28% for complex cases [11]. Our patient fulfilled three out of the four key criteria for a complex gastroschisis which were atresia, volvulus and necrotic enterocolitis. Since there is a significant correlation between complex gastroschisis and the development of a short bowel syndrome, which in turn has a major impact on the overall health of the affected children, the potential complications in our case increased significantly [8,9]. Children who develop a short bowel syndrome have a mortality rate of up to 35% within the first five years. In our case, not only a short-bowel syndrome, but even an ultra-short bowel syndrome was present, since the length measurement normally takes place only from the Treitz band, which was not available because of the incomplete development.

Since the condition of our patient was so drastic, we assumed that without adequate therapy the patient would have no chance to survive permanently. Although there has been a long trend away from small bowel transplants in literature [12] and promising results for STEP in children with ultra-short bowel syndrome with autologous intestinal reconstruction surgery, evidence was only given for small case numbers [13,14]. Compared to our case, the average time to STEP was 108 days after birth, and patients in other studies showed greater length of the small intestine [15,16]. Nevertheless we decided to do the STEP after 37 days despite some uncertainty, which resulted in a total of nine surgical procedures over a 357-day period to treat the gastroschisis and the USBS. In such a clinical picture, one should not underestimate the number of surgical interventions that may occur in the course of the patient's first year. It is definitely something to weigh the risks with.

On the one hand, our project was favored by the extensive prenatal counseling of the parents in our clinic, which took place even before birth. We informed the parents about possible consequences and associated therapeutic options of complex gastroschisis. On the other hand, we began the treatment with a planned cesarean section. Although there seems to be no remarkable advantage in literature [17], it is usually recommended to do a planned cesarean section around the 38th week of pregnancy in Germany. The aim is to prevent an early birth of the newborn, which could lead to potential formation of membranes and adhesions of the intestinal wall.

In addition, the present conditions with the dilation of the small intestine and the present ileus were beneficial for the project by STEP, which facilitated our decision at that moment, because a too small diameter of the small intestine is a limitation for performing a STEP. If that would have been the case, one could have thought about a surgical treatment with Bianchi procedure. The strong adhesions of the intestine were not critical to the choice of therapy, but required the surgeon's skills during the surgical process. Whether the therapy with such an early STEP is basically correct or not, can probably only be said if related cases are treated with a similar procedure in the future. In our case, the courage to opt for early therapy was rewarded, and in a comparable case we can clearly recommend our approach.

Conclusion

To summarize, to the best of our knowledge, the present case report is the only case so far with such an early and documented combined therapy of a gastroschisis with accompanying ultra-short bowel syndrome. Although the serial transverse enteroplasty procedure is a recognized therapy for ultra-short or short bowel syndrome, it is usually not performed that early after birth. We were able to show that a technically clean implementation of the early STEP procedure can have a positive impact on the course of a child's illness despite high operational burden. The concomitant symptoms in this case could be attributed to an infected Hickman catheter, as well as to the ileus resulting from the short bowel syndrome. There is therefore no increased risk of infection or complications due to such an early operation, but it should still be clarified in comparison with other cases. We recommend our surgical procedure which not only treats gastroschisis, but also an accompanying ultrashort bowel syndrome. Based on the scarce information on possible strategies for neonatal surgery with gastroschisis and ultra-short bowel syndrome, we have decided to publish our experience in treating this condition.

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