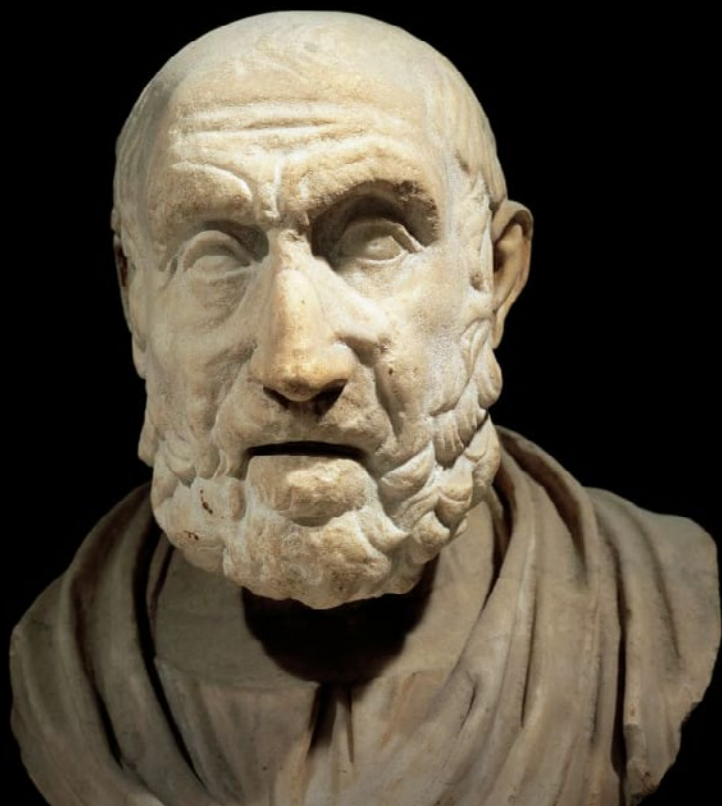


SWISS SOCIETY OF NEONATOLOGY

Irreversible intestinal failure:
how would you decide?

September 2019



INTRODUCTION

This case was presented as the Ethical Case of the Day at the 5th World Congress on Pediatric Critical Care in Geneva, Switzerland, more than 10 years ago. While progress in medicine has undoubtedly taken place in the meantime, ethical challenges illustrated by this case still occur in neonatal and pediatric intensive care and remain difficult to cope with.

CASE REPORT

This female infant was born to a 25-year-old G1/P1 from Sri Lanka at 34 1/7 weeks of gestation. Pregnancy had been uncomplicated with normal serologies and ultrasound examinations. The mother was admitted with preterm labor and shortly thereafter delivered her baby by normal vaginal delivery. The placenta was noted to be small with scattered calcifications.

The preterm infant adapted well with Apgar scores of 8, 9, and 10 at 1, 5, and 10 minutes, respectively. Arterial and venous umbilical cord pH values were 7.30 and 7.40. There was symmetric growth restriction with a birth weight of 1500 g (P3 – 10). Mild abdominal distension was the only abnormality observed in the delivery room. A nasogastric tube was placed, a 10 % glucose solution was started, and the infant was admitted to the NICU.

When normal volumes of enteral feedings were initiated on the first day of life (20 ml / kg / day, i.e. 4 ml 3 hourly), large gastric residuals (10–20 ml) were noted. A babygram was obtained and showed several dilated loops of bowel and absence of rectal gas (Fig. 1).

On ultrasound examination of the abdomen, there was no evidence of volvulus. Rectal irrigation with normal saline was unsuccessful. At this point, the parents were informed via an interpreter that surgery would be necessary to determine the cause of bowel obstruction.



Fig. 1

Babygram on DOL 1: several loops of massively dilated bowel and absence of gas in the rectum.

Laparotomy was performed on DOL 3 and revealed multiple intestinal atresias affecting both small and large bowel (i.e., intestinal atresia Grosfeld type IV); in addition, malrotation was noted (Fig. 2). The surgeons resected the first atretic part (34 cm distal to the pylorus), performed an end-to-end anastomosis and constructed a double lumen jejunostomy (Fig. 3). A Broviac catheter was placed at the same time for anticipated long-term parenteral nutrition.

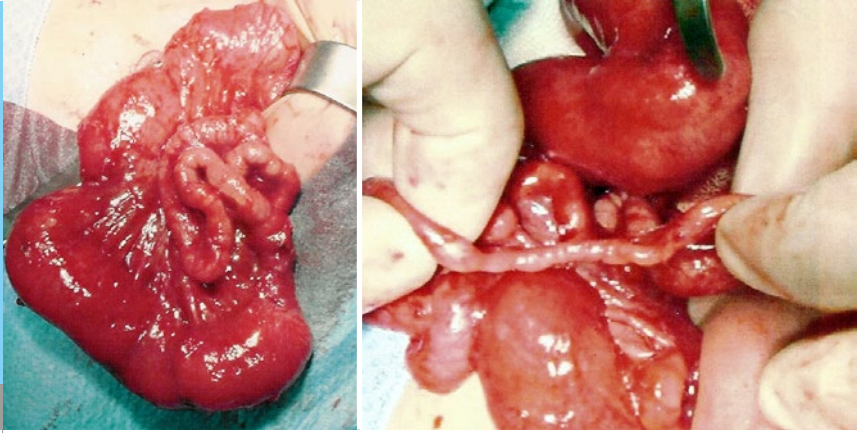


Fig. 2

On DOL 3, laparotomy was performed and revealed multiple bowel atresias.

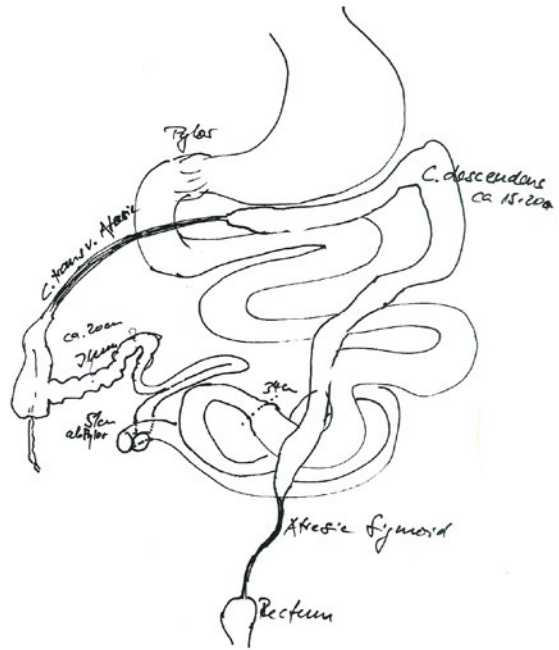


Fig. 3

Surgeon's drawing of the operative findings on DOL 3 (Dr. H. Winiker).

Postoperatively, large gastric residuals persisted, suggesting gastric outlet obstruction (Fig. 4). In addition, there was only minimal output from the jejunostomy. The parents were informed that revision laparotomy was required, and that the prognosis of their child would depend on the length of salvageable bowel. On DOL 13, a 2nd laparotomy was performed with adhesiolysis and revision of the jejunostomy.



Fig. 4

Abdominal X-ray on DOL 11: dilated stomach and no air in small or large bowel.

Unfortunately, enteral nutrition was still not tolerated, and the baby remained dependent on (near) total parenteral nutrition (Fig. 5). On DOL 27, the baby was again operated: an antral web was resected and a pyloroplasty was performed; in addition, non-functional small bowel was resected, a transversostomy was constructed (Fig. 6) and a liver biopsy was obtained. Unfortunately, gastrointestinal tract function did not recover (Fig. 7).

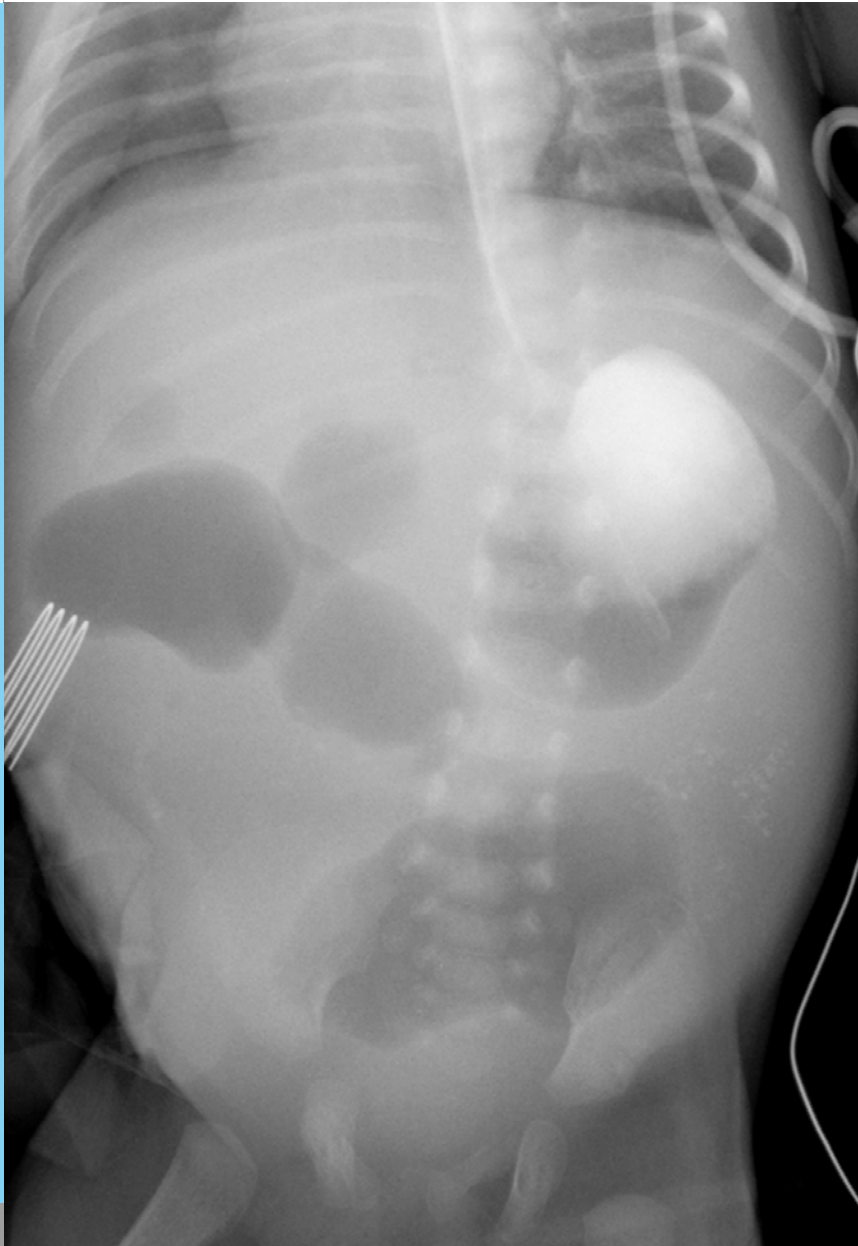


Fig. 5

Abdominal X-ray on DOL 22: persistence of both dilated stomach and dilated loops of bowel.



Fig. 6

Surgeon's drawing of the operative findings during the 3rd laparotomy on DOL 27 (Dr. H. Winiker).

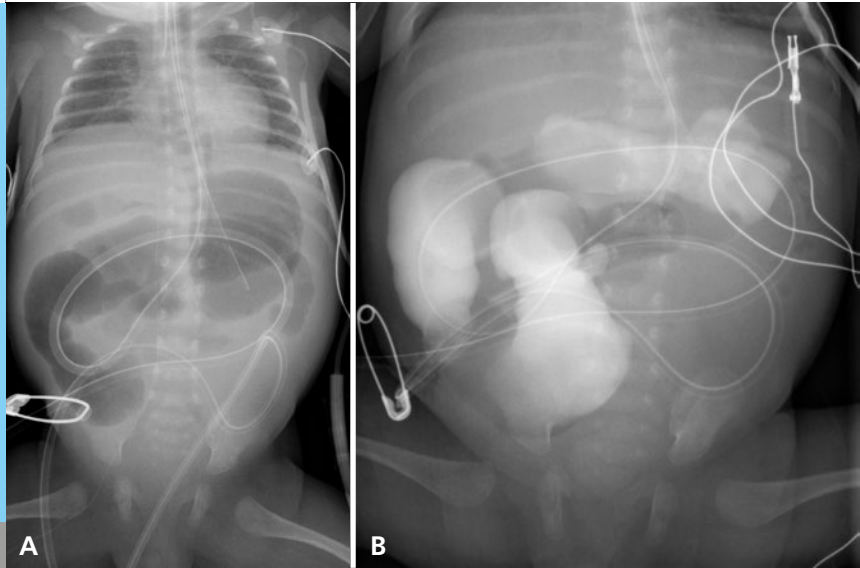


Fig. 7 A

B

*Abdominal X-ray on DOL 28 (A) and DOL 35 (B):
persistence of dilated loops of bowel.*

To decompress the proximal short bowel, a large bore tube was inserted through the jejunostomy and advanced into the proximal short bowel. Nutrition tolerance, however, did not improve (Fig. 8).



Fig. 8

DOL 48: Insertion of a large bore tube to decompress the proximal short bowel.

Parallel to the surgical interventions, numerous episodes of catheter-associated infections had to be treated with multiple courses of antibiotics and replacement of central venous catheters. Progressive cholestasis with elevated liver enzymes was found to be related to CMV hepatitis (Fig. 9) and/or long-term parenteral nutrition.

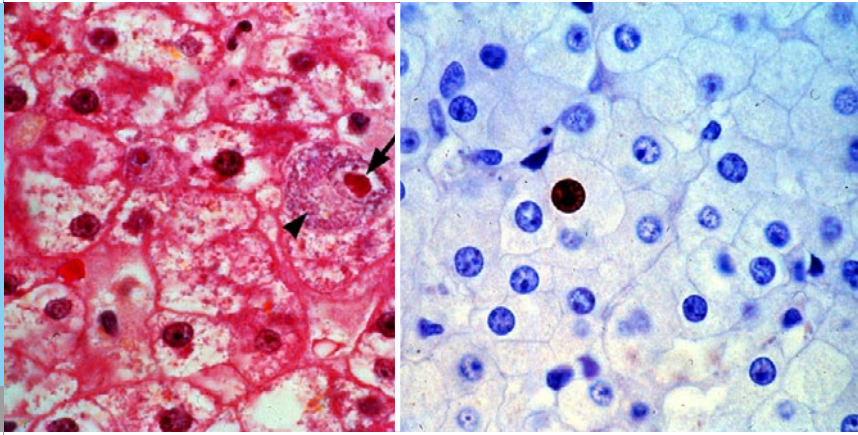


Fig. 9

Liver biopsy on DOL 68: evidence of CMV hepatitis.

The parents were informed that there was no bowel function, and that the liver was affected by a viral infection; the latter would now be treated with an antiviral drug (ganciclovir for 6 weeks). Unfortunately, cholestasis persisted, and, at 3 months of life, there was recurrent vomiting and persistence of large jejunostomy losses (250–350 ml/day), regardless of the type of nutrition used (breast milk, various formulas). Attempts to condition the distal small and large bowel were unsuccessful and increasingly painful.

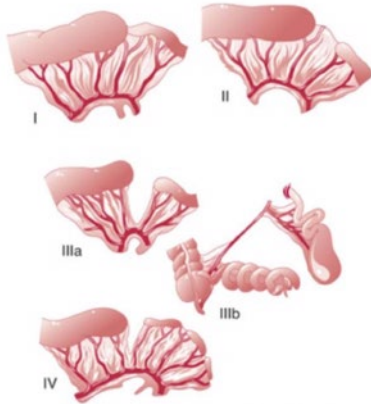
Liver and small bowel transplantation was discussed with two centers in Europe, but eventually not considered to be a viable option. The parents (refugees from Sri Lanka) and their extended family (including an uncle who was a physician) were informed accordingly.

The patient was increasingly uncomfortable. All manipulations appeared to be painful. Nausea and vomiting, febrile episodes, poor weight gain and slow psychomotor development persisted over weeks. Finally, there was a unanimous decision to redirect care. Comfort care was provided with tender loving care, analgesia and sedation. Parenteral nutrition and hydration were both discontinued. Only 36 hours later, the patient died at an age of 4.5 months.

DISCUSSION

This section will only briefly discuss different forms of intestinal atresia and then focus on the ethical aspects of this case and issues surrounding redirection of care. Finally, more recent data on the outcome of ultrashort bowel syndrome in infancy will be presented.

Approximately 95 % of intestinal obstructions diagnosed in the first 2 weeks of life are due to atresia and/or stenosis of the small intestine (1, 2). Atresia is more common than stenosis and represents complete luminal obstruction of a hollow viscus, whereas stenosis involves partial occlusion. The incidence of atresia of the small bowel ranges from 1 case in 332 live births (3) to 1 case in 5000 live births (4). No specific racial or sex predilection is known. Grosfeld et al. described the spectrum of intestinal atresias and described a classification system (Fig. 10) (5).



- Type I:** stenosis or mucosal web
- Type II:** fibrous cord between two bowel ends
- Type IIIa:** blind-ending proximal and distal bowel loops with a V-shaped mesenteric defect
- Type IIIb:** apple peel deformity (blind-ending proximal jejunum, absence of a large portion of the midgut, and a terminal ileum that is coiled around its ileocolic blood supply)
- Type IV:** multiple atresias of any kind

Fig. 10

Grosfeld classification of intestinal atresias:

(source: www.slideplayer.com)

More than 10 years ago, after extensive surgical interventions, our patient's long-term prognosis was judged to be extremely limited. Therefore, when the patient's burden was weighed against the potential benefit, redirection of care was unanimously considered to be appropriate. At this point, discussions focused on what exactly contributes to comfort care. While the provision of tender loving care, appropriate analgesia and sedation was undisputed, withholding parenteral nutrition or parenteral hydration raised concerns among health care professionals. The decision to stop the administration of any enteral fluids was made since this was associated with vomiting and obvious abdominal discomfort. Furthermore, the provision of parenteral fluids was felt to be optional, unless the infant would display clear signs of thirst. Once all means of artificial fluid and nutrition supplementation had been discontinued, the infant rapidly died. Analgesia and sedation were provided as needed, and parents and health care professionals perceived no obvious suffering.

Experience with withholding of artificial nutrition and hydration (WANH) in neonates is still limited. The first case of this concept was published in 1988 when parents of a severely asphyxiated term infant asked health care providers to consider WANH to allow their son to die (6). In a study from Canada, Hellmann et al. from the Hospital for Sick Children in Toronto reported their experience with WANH in 15 neonates

(representing 5.5 % of all deaths and 0.5 % of all admissions) (6). The median (range) gestational age was 40 weeks (31–42) and birth weight was 3409 g (2000–4640 g). The reason for WANH was predicted poor outcome due to severe neurological injury/disease. The median (range) time between WANH and death was 16 days (2–37 days). All parents reported favorable perceptions of this end-of-life care practice. The authors concluded that “WANH can be a tenable, justifiable and humane practice in the NICU.” (6).

In 2017, a review article summarized more recent experiences with ultrashort bowel syndrome in infancy. The authors noted that there had been considerable advances in the management of intestinal failure over the past 10–15 years. Survival probabilities on prolonged parenteral nutrition at 2, 5, 10 and 15 years had then reached 97 %, 89 %, 81 % and 72 % (7). While long-term parenteral nutrition is the mainstay of treatment, promotion of gut adaptation is vital to improving long-term survival. The latter can be achieved by optimizing feeds, reducing intestinal failure associated liver disease (IFALD) and catheter-related bloodstream infections. The authors argued that only a small proportion of infants with ultrashort bowel syndrome would require intestinal transplantation (7). Despite these advances in medicine, a subgroup of infants will still have less favorable prognoses, and careful planning and execution of appropriate end-of-life care, as in our case, will remain the only option.

REFERENCES

1. Grosfeld JL, Clatworthy HW. The nature of ileal atresia due to intrauterine intussusception. *Arch Surg* 1970;100:714–717 (no abstract available)
2. Singh V, Pathak M. Congenital neonatal intestinal obstruction: retrospective analysis at tertiary care hospital. *J Neonatal Surg* 2016;5:49 (*Abstract*)
3. Walker WA. Congenital anomalies. In: Walker WA, Hamilton JR, Watkins JB, Durie PR, Walker-Smith JA, eds. *Pediatric gastrointestinal disease*. 3rd ed. London: BC Decker, 2000: 556–561 (no abstract available)
4. Okada PJ, Hicks B. Neonatal surgical emergencies. *Clin Pediatr Emerg Med* 2002;3:3–13 (no abstract available)
5. Grosfeld JL. Operative management of intestinal atresia based on pathologic findings. *J Pediatr Surg* 1979;14:368–375 (no abstract available)
6. Miraie ED. Withholding nutrition from seriously ill newborn infants: a parent's perspective. *J Pediatr* 1988;113:262–265 (no abstract available)
7. Hellmann J, Williams C, Ives-Baine L, Shah PS. Withdrawal of artificial nutrition and hydration in the neonatal intensive care unit: parental perspectives. *Arch Dis Child Fetal Neonatal Ed* 2013;98:F21–F25 (*Abstract*)
8. Batra A, Keys SC, Johnson MJ, Wheeler RA, Beattie RM. Epidemiology, management and outcome of ultrashort bowel syndrome in infancy. *Arch Dis Child Fetal Neonatal Ed* 2017;102:F551–F556 (*Abstract*)

SUPPORTED BY



CONTACT

Swiss Society of Neonatology
www.neonet.ch
webmaster@neonet.ch