REVIEW ARTICLE

Chronic Intestinal Failure in Children

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SUMMARY

Background: Chronic intestinal failure (CIF) in childhood is caused by congenital malformations and inflammatory diseases of the gut. Its reported prevalence is 13.7 per million population. Long-term home parenteral nutrition has dramatically improved the life expectancy and quality of life of children with CIF. The affected children are now treated with parenteral nutrition at home as soon as their medical state and family circumstances allow.

Methods: The authors present data from a patient registry and review publications retrieved by a selective literature search.

Results and conclusion: Children with CIF can now be expected to survive beyond adolescence, at the very least, and enjoy good quality of life. This goal can only be achieved if nutritional therapy is carried out safely and the affected children's development is closely monitored by an interdisciplinary team that consists of primary care physicians/family doctors, neonatologists, pediatric gastroenterologists, and pediatric surgeons. Moreover, the prevention, early detection, and appropriate treatment of complications such as infection, liver disease, renal dysfunction, and disturbances of bone metabolism is of vital importance. The patients' families must be supported by specially qualified ambulatory nurses and social workers. Treatment with parenteral, enteral, and oral nutrition and surgery enables most infants with CIF to meet all their nutritional needs orally by the time they start going to school. For children who suffer from intractable complications, intestinal transplantation provides a real and increasing chance of survival.

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The term “chronic intestinal failure” (CIF) refers to the body's inability to meet its energy and nutritional needs through the gastrointestinal tract (1). In pediatrics, a number of diseases of the gastrointestinal tract, both congenital and acquired, acute and chronic, lead to CIF. Malformations, infections (e.g. necrotizing enterocolitis), and blood circulation disorders with gastroschisis and volvulus lead to short bowel syndrome. Functional causes include malabsorption, e.g. microvillous atrophy, and transit disorders such as aganglionosis (2). Prevalence can only be estimated on the basis of registry data (*see Table 1*). A UK center reports a point prevalence of 13.7 per 1 000 000 children (3). There is no internationally agreed definition. In Italy, 87 of 100 000 live births are diagnosed with CIF according to the definition used there (4). In Germany, period prevalence of CIF in children is probably underestimated at 170 per year. From 2000 to 2010, data were gathered on 235 children and adolescents with CIF on a voluntary basis at the Institute for Nutritional Sciences, Justus Liebig University Gießen, Germany (5). The number of children not included is unknown; 50% were reported from 9 centers (*Figure 1*).

High risks but good chances

The treatment of children and adolescents with chronic intestinal failure poses a great challenge. This is because of its clinical picture, which is usually complex. Simple though the use of parenteral nutrition may seem at first, particularly as standardization increases, the requirements for long-term nutrition management, nursing care, and the prevention of complications that continue to cause avoidable deaths are complex. There are no figures available on this, because chronic intestinal failure is not mentioned as a cause of death. Alongside primary nutritional intake and nutritional transport disorders there may also be secondary complications, e.g. growth disorders, bacterial miscolonization of the intestine, chronic diarrhea, infections, and liver disease (*Box*). An interdisciplinary treatment team with primary care provided by pediatricians and family doctors, experienced pediatric gastroenterologists and pediatric surgeons, and a specialized nutrition team can prevent complications and where necessary identify them early. This team also includes providers of social, psychological, and teaching-based care, in order to provide tailor-made treatment for the various strains faced by the whole family.

By no means simple, however, are reliable care for patients in the form of infusions and equipment, and

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TABLE 1

Most common causes of intestinal failure in children^{*1}

Disease	ITR data	German registry
Gastroschisis	21%	47%
Volvulus	18%	
Necrotizing enterocolitis (NEC)	12%	
Intestinal atresia	7%	
Chronic intestinal pseudoobstruction (CIPO)	9%	23%
Hirschsprung's disease and aganglionosis	7%	6%
Microvillous inclusion disease (MVID)	6%	
Other malabsorption, e.g. tufting enteropathy	4%	24%
Other causes of short bowel syndrome	4%	
Other motility disorders	2%	
Other causes, e.g. intestinal infarction/thrombosis	2%	
Tumors	1%	
Repeat transplantation	7%	

^{*1} ITR: Intestinal Transplant Registry; German registry: database of the University of Gießen

educational and nursing support for parents for home parenteral nutrition. The latter must be begun as early as possible.

The aim of treatment: intestinal rehabilitation and normal growth

The intestines of both premature and full-term babies are very adaptable: Neonates can achieve complete intestinal rehabilitation with intestinal lengths as short as 25 cm and, even in intestinal lengths <25 cm, children have a good prognosis regarding quality of life and life expectancy (6). Advances in neonatal care, pediatric intensive care, pediatric surgery, and especially parenteral nutrition have significantly improved the prognosis of children with CIF. One-year survival rates of 97% and five-year survival rates of 89% are reported (7). Patients receiving long-term parenteral nutrition in the German-speaking world received parenteral nutrition for a median of 2.8 years (mean 5.2 years, minimum 90 days, maximum 22 years); most achieved complete intestinal adaptation. This is in line with the experience of the Center for Long-Term Parenteral Nutrition in Children in Paris (8) (Figure 2).

It is important that doctors and parents who are faced with the challenges of home parenteral nutrition are aware of this prognosis. Parents have joined together in an association called *Kinder in schwieriger Ernährungssituation*, literally meaning “Children with Difficult Nutritional Status” and abbreviated *KiSE* (website www.kise.de, in German only). They provide each other with mutual support.

The overriding aim of treatment is normal growth and intestinal rehabilitation. All oral and enteral

nutrition options involving special diets, surgery, and drugs are used to achieve this aim. Figure 3 shows the development of the weight of children receiving parenteral nutrition. It remains unclear whether thriving in the first ten years of life is due to better treatment during this period or whether in the second ten years of life only those—more seriously affected—children remain in the cohort who have failed to achieve enteral adaptation earlier.

This article aims to describe a rare, complex clinical picture and its treatment. Treatment success requires intensive interdisciplinary cooperation.

Data from a patient registry, the authors' experience, and the results of a selective search of the literature for “intestinal failure in children” in PubMed covering the period 2007 to 2011 are reported.

Intestinal adaptation involving oral/enteral nutrition alone

Because chronic intestinal failure in children is rare, there are only a few review articles and collections of case histories, alongside descriptions of individual cases (1, 6, 11, 13, 20). Following intestinal resection, nutrition via the intestinal lumen should be started as soon as possible, in other words during the initial phase of diarrhea and gastric hypersecretion. Intestinal passage must be continuous, wherever possible from the mouth to the anus, but if necessary to a stoma. Even if no nutritional benefit can be expected from oral or enteral intake, the intake of nutrition does stimulate the mucous membranes, gut hormone production, and the formation of physiological flora; this leads to regeneration of the mucous membranes and encourages bile flow (9). Intestinal adaptation via mucous membrane hyperplasia and muscle hyperplasia is associated with dilation and elongation of the small intestine; resorptive performance can increase up to full enteral adaptation. Gut hormones, growth factors, and digestive secretions stimulate intestinal growth (10).

Oral or enteral nutrition must be selected on an individual basis; there are no dietary restrictions unless there are specific digestive problems. Complex carbohydrates, proteins, and fats reduce osmolality, fluid loss through diarrhea, and abdominal complaints.

Solid food should be introduced in line with the patient's age, in order to prevent the development of food aversion (11). Some patients benefit from continuous enteral probe feeding; for others, gastric bolus feeding is superior.

Glutamine and growth hormone treatment have been researched in adults (12), but to date there is no reliable evidence of the benefit of glutamine, growth hormone, or medium-chain triglycerides (13). Initial reports on the use of glucagon-like peptide (GLP-2) are promising (14).

The need for interdisciplinary cooperation

In both our experience and that of others (15–19), it is vital for the successful treatment of CIF and for children's and adolescents' life expectancy and quality

of life that the various tiers of treatment (enteral and parenteral nutrition, surgery) are well coordinated, so that tailor-made treatment can be developed and jointly implemented. Swift decisions and consistent action can prevent nutritional deficiencies, the risk of infections, and growth disorders, as well as the onset of a liver disease. Although there is no statistical evidence, the rarity of chronic intestinal failure in children and treatment results from large treatment centers, outside Germany (in Paris, Birmingham, Toronto, and Boston), suggest that the necessary experience can only be gathered at centers. Many children in Germany are currently transferred to a treatment center after too long a latency period: Specialized centers tend to be too little-known, German centers' activities and facilities cannot be compared to those of centers in other countries because of a less consistent referral behaviour of peripheral institutions, and risks are often underestimated until irreversible damage has occurred.

The range of outpatient care facilities on offer from providers of solutions for infusion and home care services, and the guidelines that are available (20, 21), tend to contribute to the impression that patients can be treated even without sufficient prior experience. However, only treatment performed at centers with sufficient experience can prevent life-threatening situations and provide a chance of intestinal rehabilitation for children with intestinal failure.

Preventing catheter-associated complications

Because of the long-term dependency on parenteral nutrition, the handling and care of venous catheters are a cornerstone of the treatment of patients with intestinal failure. Single-lumen Broviac or Hickman catheters with a subcutaneous tunnel at the anterior chest wall are

FIGURE 1

Distribution of patients in the Gießen registry for children and adolescents receiving home parenteral nutrition among the 114 outpatient facilities and practices involved. 57 of these outpatient facilities and practices treat a maximum of 4 patients each, while 9 centers treat an average of 12 children each

preferred; the tip lies at the junction between the sinus of the vena cava and the right atrium. They are used for feeding only. Every additional lumen increases the risk of infection (22). Of the children included in the database, 88% have a tunneled catheter, and only 9% a port. Ports promote deposit formation, because the dominant flow inside them is nonlaminar. In the patients included in the research in Gießen, catheters were worn for a median of 415 days (maximum 4883 days, standard deviation 873 days). In a prospective cohort study conducted in adult patients with CIF who were receiving home parenteral nutrition, approximately two catheter-associated complications (one of which was an infection) occurred for every 1000 catheter days (23). The

BOX

Risks and complications associated with short bowel syndrome and/or chronic intestinal failure

- Reduced growth
- Metabolic disorders:
 - Acidosis, D-lactic acidosis
 - Fluid and electrolyte metabolism disorder
- Osteopathy
- Nephropathy
 - Hyperoxaluria
 - Kidney stones
 - Kidney failure
- Peptic disorders
 - Ulcer
 - Gastroesophageal reflux
- Intestinal complications
 - Stenosis with prestenotic dilation
 - Pseudoobstruction
 - Intestinal obstruction
 - Prolapse, stenosis, or infection of the stoma
 - Excessive bacterial growth in the small intestine
 - Bacterial translocation
 - Secretory and osmotic diarrhea
- Problems with venous access
 - Obstruction, dislocation, or damage of the catheter
 - Thrombosis
 - Catheter sepsis
- Liver disease
 - Cholestasis, steatosis, fibrosis, and cirrhosis of the liver
 - Gallstones, cholecystitis, biliary sludge
- Psychosocial problems
 - Hospitalism
 - Food aversion

Endpoints of 44 children and adolescents with chronic intestinal failure receiving parenteral nutrition according to data from the Gießen registry (no transplantation; deaths caused by infection, liver failure, and other causes)

FIGURE 2

data of the Gießen registry record 2.8 infections per 1000 catheter days, 1.5 per 1000 resulting in a change of the central venous catheter. This order of magnitude also applies to other pediatric registries (24).

Preventing liver disease

A British study of adults with CIF showed that cooperation between centers and local hospitals in performing home parenteral nutrition can reduce the complication rate to a lower level (25). The example of a liver disease associated with intestinal failure shows how essential prompt cooperation is. Transaminase levels increase in 40% to 60% of patients with CIF who receive long-term parenteral nutrition. In children, this liver disease most commonly takes the form of cholestasis and fibrosis but sometimes manifests as fatty liver; it is caused by multiple factors (26). If there are also other factors, such as intestinal inflammation, the risk of rapid liver damage increases. These processes must

be identified early and promptly. Only swift, consistent treatment in consultation with experienced pediatric hepatogastroenterology specialists improves the prognosis (Figure 3). Preventing hypercaloric nutrition involving more than 100 kcal/kg bodyweight per day, intermittent parenteral nutrition with cyclic release of gastrointestinal hormones, and early oral or gastric feeding contribute to this (27). Risks of recurrent infections, from phytosterol-rich lipid emulsions, high vitamin A and low vitamin E intake, and hepatotoxic drugs remain. The use of lipid emulsions containing fish oils that have an anti-inflammatory effect thanks to omega-3 fatty acids shows success in the early treatment of liver disease in small, uncontrolled case series (28).

The most important contribution to the prevention and treatment of liver diseases is provided by intestinal rehabilitation that allows intravenous intake of glucose and lipids to be reduced below the relatively safe boundary of 7 or 1 g/kg bodyweight (29). Experience from Canada shows that the treatment of patients in a pediatric short bowel syndrome center based on clear guidelines on parenteral nutrition, swift weaning off parenteral feeding, and aggressive treatment of complications reduces the incidence of liver diseases and increases the survival rate (30).

Surgery is challenging

Short bowel syndrome also presents a major challenge for surgical treatment. Optimum treatment is provided only when all issues are considered. Surgery is not limited to stenosis removal or intestinal lengthening. When and to what extent resection of the intestine is necessary is decided during the neonatal period. Primary complete resection is only performed in clear cases of necrosis; if some blood circulation remains, the extent of resection can be reduced by enterostomy and two-step surgery. The declared goal must be to

Development of weight of patients in the Gießen registry for children and adolescents receiving home parenteral nutrition

FIGURE 3

Figure 4: Surgical illustrations of BIANCHI intestinal duplication according to Hosie S, Wessel L, Waag K-L: Darmdoppelung nach Bianchi als chirurgische Therapie des Kurzdarmsyndroms. *Ellipse* (1998) 14: 61–64

preserve as much of the resorption surface of the intestine (including the colon) as possible. The aim following successful ileostomy is early restoration of intestinal continuity to make early breastfeeding and age-appropriate, complex nutrition possible and prevent the loss of large quantities of fluid.

CIF surgery aims to remove obstructions, establish normal chyme transport, remove intestinal miscolonization, and achieve intestinal adaptation. Bianchi longitudinal intestinal lengthening and tailoring (LILT) (31) or transverse intestinal lengthening using a serial transversal enterostomy procedure (STEP) (32) reduce reactive dilation of the intestine and improve contact time between chyme and the intestinal surface, thereby also improving resorption capacity (*Figure 4*). Surgical intestinal lengthening requires sufficient dilation of the small intestine. Indications are dependence on parenteral nutrition with insufficient resorption capacity and recurrent bacterial miscolonization.

Successful intestinal lengthening, defined as attainment of intestinal autonomy, is achieved in approximately 70% of cases (33). Partial success means that parenteral nutrition can be substantially reduced (infusion-free days or volume of infusion reduced by more than 50%). Complications may be surgical (inadequate suturing, circulation disorders, stenosis), functional (motility disorders, redilation), or infection-related (persistent miscolonization, Crohn's-like disease). They can prolong dependence on parenteral nutrition and prevent intestinal autonomy.

Intestinal transplantation as last resort

Nowadays, children in whom no enteral feeding or weaning can be achieved despite the use of all treatment options and whose chances of parenteral nutrition are irreversibly restricted due to repeat septicemia and vascular thromboses can also benefit from intestinal transplantation. A decision regarding intestinal transplantation must not be postponed for too long.

If there is a serious liver disorder, intestinal transplantation is combined with liver transplantation. They need not necessarily come from the same donor. To date approximately 1300 transplants have been performed worldwide, 60% of them in pediatric patients

(34). In Britain it is estimated that 2 to 3 individuals per million require intestinal transplantation every year, half of whom are children. Because according to the Edinburgh center's data up to 50% of children with chronic intestinal failure after infancy meet the criteria for indication of intestinal transplantation, the actual figure may be higher (35). In 1987, when the first successful multivisceral transplantation was performed in Pittsburgh (PA), in a child, and in 1988, when the first intestinal transplant in a child was performed at Kiel University Hospital, the one-year survival rate for transplants was only 30% and the three-year survival rate only 20%. Thanks to modern induction immunosuppression therapy and long-term tacrolimus

Figure 5: Child with chronic intestinal failure at a kindergarten with stoma bag and parenteral nutrition in backpack (photo: Andreas Reeg, with permission)

immunosuppression, according to international registry data the mean five-year survival rate for patients and transplants is now between 50% and 80%, depending on the underlying disease and presurgical morbidity (36).

German transplant centers also report similar results (37). In 2009, for the first time in Germany, a child with complete aganglionosis successfully underwent transplant surgery in Tübingen. Immunosuppression therapy requires high expenditure on the detection and treatment of rejection, infections, and post-transplant lymphoproliferative disease (PTLD), which occurs in up to 30% of children (32).

In addition to the underlying disease, the prognosis of intestinal transplantation also depends on the frequency of previous surgery, liver function, comorbidity, and nutritional status. Selection of the correct time for transplantation is a decisive factor, because both patients waiting for a combined liver and intestine transplant as result of liver disease and recipients who are hospitalized before transplantation have a significantly worse prognosis (32). The advances over recent decades in the treatment of children with chronic intestinal failure were made possible, above all, by improvements in parenteral nutrition, surgical techniques, and immunosuppression therapy following transplantation.

Conclusion

In order to optimize enteral nutrition and develop medication strategies to promote adaptation and prevent liver disease and generate new surgical techniques, large-scale, multicenter studies are required. For children with chronic intestinal failure in Germany there are already centers with the treatment capacity and experience as well as the necessary specializations to provide the whole range of treatment from early intestinal adaptation to intestinal transplant. However, these treatment centers must be equipped in a way that

takes into account the staff-intensive nature of complex treatment of children with intestinal failure and allows for appropriate quality management. Early contact with one of these centers and interdisciplinary treatment with the aim of intestinal rehabilitation, including all physicians and the multidisciplinary team, is a decisive factor for establishing digestive function and preventing secondary complications (*Figure 5*).

Conflict of interest statement

Dr. Kohl has received consultancy fees and fees for preparing scientific continuing education events from Baxter. He has also received monies paid into a third-party account for a research project he himself initiated from Fresenius.

Dr. Busch has received lecture fees from Fresenius-Kabi.

Prof. Krawinkel has received consultancy fees from Fresenius-Kabi. He has received fees for preparing scientific continuing education events from Caremark and Fresenius-Kabi. He has also received monies for a research project he himself initiated from both companies.

Prof. Zimmer, Prof. Wessel, and Dr. Scholz declare that no conflict of interest exists.

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KEY MESSAGES

- Neonates with intestinal failure have a good chance of survival if they receive parenteral nutrition promptly and consistently.
- The aim of treatment is intestinal rehabilitation that enables children to become independent of parenteral nutrition.
- Successful treatment requires the close collaboration of pediatricians and pediatric surgeons and the benefit of experience at specialized centers.
- Liver diseases and infections can be prevented if complications that develop are treated promptly and consistently.
- For children who do not achieve intestinal adaptation, there are surgical procedures available to lengthen the intestine and improve resorption and motility, as well as the option of intestinal transplantation.

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