

Tube weaning according to the Graz Model in two children with Alagille syndrome*

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Abstract: Two children were weaned from long-term tube feeding after liver transplant because of Alagille syndrome. The children were successfully weaned, one in seven days and the other in 13 days, using our standard and highly specialized intensive treatment protocol. Normal feeding behavior and stabilization of body weight were established. Children fed by long-term enteral tubes can be weaned from enteral feeding even after a long period of treatment. The return to age-appropriate self-feeding should be introduced as early as possible. Our weaning program time is brief and effective and can be recommended generally to improve quality of life and withhold unintended side-effects of enteral nutrition.

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Ongoing improvement in pediatric liver transplantation has been seen over the last years. Such improvement allows careful attention to be directed toward understanding and alleviating the psychosocial risks or side-effects of tube feeding, as well as more direct focus on quality of life after transplantation. Children fed by tube are at a risk to suffer from complications and medical and psychosocial unintended side-effects, such as gastroesophageal reflux (GER), dumping syndrome, and problems in interaction with peers and parents (1). Long-term enteral nutrition is often associated with infections or digestion dysfunction. However, exclusive enteral nutrition is a burden for the children in contrast to normal eating behavior (2). It is clearly shown that tube feeding does increase the mortality rate in children (3). To improve the quality of life of these children, the Graz tube weaning model has been developed and published in German (4, 5) and English (6). In the last seven years, working according to this protocol more than 156 infants from various

European and Mediterranean countries have been treated. About 90% of these children could be weaned successfully (7). Therefore, a highly specialized, motivated, and competent team is needed.

We present two children fed by tube (8), who were weaned successfully. These case reports show which subdisciplines and therapies are involved in the inpatient weaning program.

As outpatient weaning models such as behavioral treatment (9) or nutritional support are not working in severely ill children, weaning must be done on hospitalization in these patients.

The aim of this case report was to show that weaning is possible even in severely ill children after liver transplantation. The treatment protocol is not specific for children after transplantation but these patients can be treated too.

Case report

Both of our patients received a liver transplantation because of Alagille syndrome (10) and

were admitted by pediatricians. In both the duration of tube feeding has been decided by their treating specialist. Both patients had several weaning trials which failed in outpatient setting. In both patients tube weaning was not started in outpatient setting by us because of the consequences of malnutrition stressed parents and medical personnel. As liver transplantations are done in Austria mainly in Innsbruck patients came for weaning to Graz (Austria) which is a 6-h drive by car.

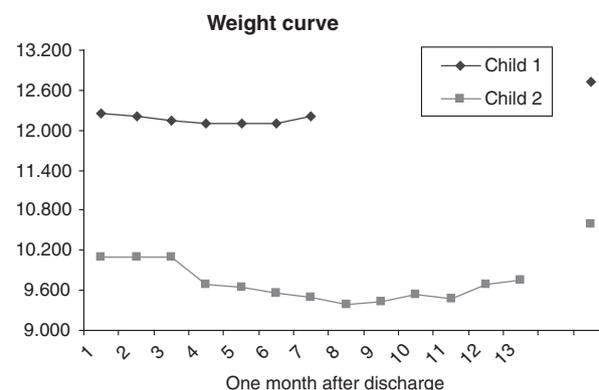
Case vignette 1

Patient 1 was born without complications in the 39th gestational week with a weight of 3790 g and a length of 52 cm. Because of increasing icterus he received a Kasai surgery at the age of 1 month and the allogene orthotropic liver transplantation at the age of 11 months. Because of his disease he developed severe failure-to-thrive (11) which caused the introduction of tube feeding. The patient was fed by nasogastric tube from the age of four months until admission. After liver transplantation the tube feeding was continued because of complications, including recurrent infections and rejection of the transplant. At the age of two years, the patient was admitted at our hospital with a weight of 12 250 g and a length of 81.5 cm (BMI 18.44; on the 30th percentile using the charts of Prader et al.) (12). Physical and neurological examination was normal.

The tube feeding was completely stopped on the first day of inpatient treatment, because he removed the tube accidentally by himself. In consent with the parents we decided to leave the child without tube. Subsequently his body weight decreased to a minimum of 12 100 g on day 4. The maximum weight loss was 150 g, which was 1.2% of body weight. Thereafter, he gained weight and was discharged after seven days with a body weight of 12 200 g. One month after discharge, the weight was 12 720 g. Weight data are shown in Fig. 1.

Case vignette 2

Patient 2 was born in the 40th gestational week. She received an orthotropic liver transplantation at the age of 2 years and 9 months. The percutaneous endoscopic gastrostomy (PEG) tube was implanted before transplantation to enhance weight before surgery. After transplantation the tube remained. When admitted to our ward at the age of 4.5 years she was still fed by tube. Her bodyweight at admission was 10 100 g and the length was 90 cm (BMI 12.5 – both



- (3) The infant should be able to swallow its saliva.
- (4) No anatomical defect in the gut passage should be present.

Additionally both parents must agree to the weaning protocol and accompany the infant through out the program. They have to learn to read and accept the infant's non-verbal cues of hunger and food refusal or avoidance and must be sensitive and respectful towards them. They must be able to notice and discuss irritations or bad feelings with members of the weaning team.

The last but probably hardest task is the necessary developmental change process in the clinical team: learning to eat can only happen in a secure and good natured environment with a high level of know-how in the field of normal eating and feeding development, starvation, malnutrition, and any condition of severe failure to thrive. The team must be available 24 h a day and should include pediatricians, laboratory personnel, surgeons, radiologists, an experienced nursing team, occupational therapists, speech therapists, dieticians, physiotherapists, developmental psychologists, and experts from psychiatry, cardiology, and gastroenterology, if needed. In some cases a psychiatrist for parents support is needed.

The standardized inpatient tube weaning program includes the following items:

- (1) Pediatricians are responsible for monitoring body weight and the infant's general medical condition, especially in the period of fast reduction of enteral nutrition in the first days of treatment. Invasive diagnostic procedures are only permitted in cases of emergency. If needed, the pediatrician should contact other specialists.
- (2) The nursing team is responsible for nursing and nurturing the child and the parents emotionally to reduce parental stress. Preparing the dishes and food should happen as a daily play picnic.
- (3) The daily play picnic: within this therapy item various food is offered, prepared suitable for children using plastic dishes. The children are allowed to play with the food and make experiences with touching and tasting new flavors. During the picnic situation parents can watch it through a one-way mirror. Children are guided by members of our team following the aim of self-regulation and not catering the child.
- (4) Video analysis is needed to identify intrusive behavior and to reflect the child-parent interactions together with the parents. This instrument is also helpful in the diagnostic assessment process and for aftercare.
- (5) Developmental psychology: therapy should detect and encourage deficits in age-appropriate eating behavior

within the daily 'playing eating' with puppets and other instruments.

- (6) Interaction-focused therapy occurs constantly as nearly 50% of the team is additionally trained in psychotherapy. The aim is to achieve a better quality in interaction, especially in eating and feeding situations, as most of the families of tube-fed children do show disturbed interactional cues especially concerning eating and feeding topics.
- (7) Psychoanalytic-oriented psychology is needed to work on traumatic events in the child's and parent's history. Often the parents' dyad has suffered because of the infant's illness.
- (8) SLP-therapy: through non-traumatic stimulation of the orofacial area the child makes positive experiences which correct the traumatic experiences which occur in this area. Most of the children are delayed in speech development.
- (9) Occupational therapy: promotes tactile cognition and sensory integration through stimulation of the vestibular system.
- (10) Physiotherapy: deals with pathological motor tone and feeding behavior and offers basal stimulation or craniosacral therapy. The frequency of therapy items differs from three to six times a week and might also include the parents.
- (11) Nutritional counselling: advises the parents on normal age-appropriate nutrition and in preparation of special diets, if needed.
- (12) Early intervention: in many cases, the outpatient early intervention services are very helpful for integrating the new situation into their every day life and organizing aftercare.

A typical time schedule for one week can be seen in Table 1.

Discussion

Advanced operative techniques in transplantation surgery and enteral feeding allow babies to survive by being fed enterally over a long period of time. The insertion of a nasogastric or PEG tube is technically easy and is rarely cause of any detailed discussion. There are no clear clinical criteria for such tube insertions. However, once the infant is stable on tube feeding, the question of who, when, and how to remove the tube is open and there exists little guidance in the literature. Long-term complications are listed in a few papers and include reports of surgical

Table 1. Example of time schedule for inpatient therapy

Time	Monday	Tuesday	Wednesday	Thursday	Friday	Saturday	Sunday
8.00–9.00	Ward round	Ward round	Ward round	Ward round	Ward round		
9.00–10.00	Physiotherapy		Physiotherapy	Physiotherapy	Physiotherapy		
10.00–11.00	Parents counseling	Logotherapy		Ergotherapy	Parents chat	Ward round	Ward round
11.00–12.00			Logotherapy		Ergotherapy		
12.00–13.00	Play picnic	Play picnic	Play picnic	Play picnic	Play picnic	Play picnic	Play picnic
13.00–14.00							
14.00–15.00	Ward round	Ward round	Ward round	Ward round	Ward round		
15.00–evening	Family activities, go for a walk,						

problems, speech delay, psychosocial problems, and unintended side-effects and problems associated with tube feeding itself like GER (14) and dumping syndrome (15). Tube weaning in infancy is one of the topics of psychosomatic medicine in pediatrics. To reduce tube-associated problems, various weaning models were developed in the past. The most popular one is behavioral treatment using flooding techniques. This treatment is able to wean about 45% of the children (9). But it is not useable in severely ill children. Thus, in our clinical experience flooding does produce post-traumatic feeding disorders. So, we developed a treatment protocol following the principle of self-regulated oral intake without using traumatizing therapy items. A paper submitted for publication (*Journal of Pediatrics*) shows in a retrospective analysis of 110 patients treated according to our therapy protocol showed, that more than 90% of the children could be weaned successfully. These weaned children (70%) were severely disabled, following the criteria of ICF (16).

The 'Graz Tube Weaning Program' is divided into three major parts: (i) the preconditions and variables on the infant; (ii) the challenge of the involved parents supporting their infant through this phase of learning; and (iii) the requirement of team preparation concerning the way of understanding the developmental issues of eating and feeding apart from the large variety of medical issues.

The return to age-appropriate self-feeding should be recommended as early as possible. The weaning program time is brief and effective and should be done generally to improve the quality of life. In the retrospective analysis of the 110 patients, the tube can be removed after eight days in median and mean inpatient treatment time is 21 days.

Our treatment costs \$864 per day when applied in inpatient therapy. Calculated with the inpatient treatment time of these two patients it costs \$6.048 for patient 1 and \$11.232 for patient 2. This seems very much in comparison with the costs of outpatient treatment as it is used in behavioral protocols. But in comparison with the costs of life-long tube feeding and possible complications and negative side-effects on behavior and development this seems cheaper for all healthcare systems which pay for therapy and transport of the family. Weaning the patients has an enormous socioeconomic effect. A study (17) calculated the economic and psychological costs for the caregivers of gastrostomy-dependent children with \$37.232 per year and \$15.004

per year for caregivers with disabled children without gastrostomy tube. The effective costs caused by gastrostomy tube feeding are \$22.228 per year, which is more than the cost of the presented treatment program.

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