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OUTCOMES OF PEDIATRIC SHORT BOWEL SYNDROME ASSOCIATED INSTESTINAL FAILURE (SBS-IF)

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Aim of the study: To analyze outcomes and prognosticators of pediatric short bowel syndrome (SBS) associated intestinal failure (IF).

Methods: All children with SBS-IF with parenteral nutrition (PN) >2 months or small intestinal resection >50% managed in our center were included. Patient characteristics, weaning off PN and survival were retrospectively analysed with Kaplan-Meier curves, log-rank test and Cox regression models.

Main results*:* In total, 73 patients with median follow-up of 5.4 (IQR 2.6-11) years and PN-duration of 11 (4.6-34) months were included. Causes of SBS were NEC (n=32), atresia (n=14), volvulus (n=13), Hirschsprung disease (HD, n=10), and isolated gastroschisis (n=4). Patients had 40 cm (29-61) or 25% (17-43) of small intestine, and 86% (57-100) of colon remaining. Six had end-jejunostomy and 35 ileocecal valve (ICV) preserved. 18 patients underwent lengthening/tapering and 6 other procedures to improve bowel function. Two HD-patients underwent intestinal transplantation (ITx) and two are listed. Overall 2- and 5-year weaning off rates without ITx were 64% (95%CI 52-75) and 74% (63-85). None of the patients with jejunostomy (all with HD) weaned off PN. Other negative prognosticators for weaning off were remaining small bowel length <20% or <30 cm, missing ileum and ICV, and <70% remaining colon, whereas NEC-patients had increased likelihood for weaning off (**Table**). Incidence of culture proven blood stream infections was 1.79/1000 PN-days. Derangements in liver biochemistry was observed in 26%, and 43% (23/54) had Metavir fibrosis stage ≥2 in liver biopsy. Six (8%) patients died and overall 20-year survival rate was 90% (95%CI 83-98). Among survivors height z-score was -1.6 (-2.5- -0.4) and weight percentile -4.0% (-12-2.0).

Conclusions: Despite reassuring long-term survival, blood stream infections, IF-associated liver disease and weaning off PN in patients with very short remaining small bowel, end-jejunostomy, PIMD and CI are still significant concerns in children with IF.

Variable	Hazard ratio	95%CI	P-value
NEC-diagnosis	2.22	1.28-3.82	0.005
HD-diagnosis	0.11	0.03-0.46	0.002
<20% small bowel	0.36	0.20-0.67	0.001
<30 cm small bowel	0.48	0.25-0.93	0.03
No ileum	0.33	0.19-0.57	<0.001
Removed ICV	0.33	0.19-0.58	<0.001
<70% colon	0.40	0.22-0.75	0.004