

A 25 YEAR REVIEW OF EPIDEMIOLOGY AND OUTCOME OF PEDIATRIC SBS IN THE NETHERLANDS

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Aim of the study: Short bowel syndrome in children remains a therapeutic challenge with high morbidity and mortality. There is a paucity of basic epidemiologic knowledge of pediatric SBS as nationwide data are lacking. Aims were to generate nationwide epidemiologic data on SBS in children and to investigate the outcome of pediatric short bowel syndrome in the Netherlands.

Methods: We retrospectively identified all pediatric patients (<17 years at time of diagnosis, remnant small bowel <25% for age) developing SBS between the 1st of June 1987 and 31st of May 2012, from all Pediatric Surgical Centers in the Netherlands. Multiple clinical parameters including age, remaining small bowel length, operations, TPN duration, hospitalization length and mortality were assessed. Ethics committee approval was obtained. Data provided are median (range) unless specified otherwise.

Main Results: We identified 217 children (124 boys, 93 girls). Age at diagnosis was 12 days (0-5110), 192/217 (88.5%) occurred in neonates. General incidence was 2.4/million children/year (1.0-4.5), with in neonates an incidence of 3.6/100 000 live births (1.5-8.3). Main etiological causes were: NEC 93/249 (37.3%), volvulus 58/249 (23.3%), intestinal atresia 50/249 (20.1%). Remaining small intestinal length was 27.8cm (0-90cm): jejunum 15cm(0-70), and ileum 2cm(0-70). Percentage small bowel length was 17.4%(0-25.4%).

130 children (59.9%) survived >1 week., 72 boys, 58 girls, age 8 days (0-5110). Remaining small bowel length was 20.1% (0.1-25.4), hospitalization 203 days (8-4712); TPN duration 181 days (9-4832). Median operations 8 per patient (1-28). Overall mortality was 27%. Between 1987-1992 mortality was 40%, between 2007-2012 it was 10%.

Conclusion: Following massive bowel loss 56% of SBS children did not survive, most deaths occurred within the first week after operation. Survivors required lengthy hospitalisation and TPN treatment. Incidence and prevalence is low; albeit with high impact on patient and hospital resources.