J Pediatr Gastroenterol Nutr. 2019 May;68(5):635-641. doi: 10.1097/MPG.0000000000002260.

Autologous Intestinal Reconstructive Surgery in the Management of Total Intestinal Aganglionosis.

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Abstract

OBJECTIVES:

Total/near total intestinal aganglionosis (TIA/NTIA) is the most uncommon and life-threatening form of Hirschsprung disease (HD). The management of TIA/NTIA is challenging and the role of autologous intestinal reconstructive (AIR) surgery is controversial. The objective is to evaluate the effectiveness of AIR in patients with TIA/NTIA.

METHODS:

Records from children affected by TIA and enrolled in the multicenter international Pediatric Intestinal Rehabilitation and Transplantation Registry were retrospectively reviewed.

RESULTS:

Fourteen patients with TIA were identified. TIA diagnosis was confirmed histologically at the median age of 14 days of life. All received a proximal decompressive jejunostomy. Two patients died, 4 patients had satisfactory stoma output with enteral tolerance without additional procedures, 8 underwent 10 AIR procedures (4 Ziegler myotomy-myectomy, 3 transposition of aganglionic ileum with or without myotomy, 2 simple tapering, 1 longitudinal lengthening and tailoring procedure with associated myotomy). AIR significantly reduced median stoma output, from 197 to 31 mL·kg·day ($P=0.0001$). The reduction was seen in all patients. In addition, AIR improved enteral tolerance in the long term in 5 of 8 patients (63%), and temporarily in 1, leading to a reduction of parenteral nutrition requirement from 100% to 70% ($P=0.0231$).

CONCLUSIONS:

AIR surgery in carefully selected patients may be useful and effective way to enhance residual bowel absorptive function and to reduce parenteral nutrition requirements. AIR and intestinal transplantation are complementary surgical tools in the complex treatment algorithm of TIA/NTIA.

PMID:
30614951
DOI:10.1097/MPG.0000000000002260