BMJ

World Journal of Pediatric Surgery

Call for Papers

Pathophysiology and Treatment of Hirschsprung Disease

Hirschsprung disease (HSCR) is a congenital disorder characterized by the absence of ganglion cells in the distal intestine, resulting in life-threatening functional bowel obstruction. While most children have disease limited to the rectosigmoid, the aganglionosis can extend further proximally, even affecting the entire intestinal tract in some cases. The condition affects 1 in 5,000 neonates and results from abnormal embryologic development of the enteric nervous system. Once the diagnosis is confirmed histologically by the absence of ganglion cells on rectal biopsy, surgical resection of the aganglionic segment is performed. Much variability exists in the operative approach and perioperative management of these patients, and outcomes are highly variable. Despite a seemingly successful operation, many children continue to experience constipation, enterocolitis, intestinal dysmotility, or varying degrees of fecal incontinence, and these can be due either to complications of the surgery or to functional defects inherent in the disease. Advancements in medical and surgical management, along with new insights into disease pathophysiology, are helping to improve outcomes for these children.

This topic collection focused on Hirschsprung disease will cover a breadth of topics, including its genetic and embryologic origin, diagnosis and treatment, long-term outcomes, and innovations on the horizon.

Topics to be covered:

- Genetic and embryologic origins of Hirschsprung disease
- Variants of Hirschsprung disease (e.g. long-segment, total intestinal, late presentation)
- Hirschsprung-associated enterocolitis
- History of surgery for Hirschsprung disease
- Current surgical approaches
- Controversies in operative management
- Histopathology
- Long term functional outcomes and quality of life
- Future therapies and innovations

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Submission Information

Please submit your manuscript before 31-May-2024 via the journal's submission system: ScholarOne Manuscripts. Please select the special issue name **'Pathophysiology and Treatment of Hirschsprung Disease'** when submitting your manuscript.

Preliminary enquiries and manuscript submissions may be sent in the first instance to the Journal Editorial Office (**wjps@zju.edu.cn**).

Before submitting, please consult the Author Guidelines for more information about the journal, manuscript types, and instructions for manuscript preparation.