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Hirschsprung Disease – Editorial

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Hirschsprung disease (HD) is one of the many congenital conditions that, despite significant advances in surgical therapies over the last 60 years, may be affected by poor long-term outcomes. The typical child with HD is diagnosed soon after birth, undergoes a pull-through of the aganglionic colon and rectum early in infancy, and then establishes bowel control at a variable stage in early childhood. However, the effectiveness of the pull-through (independent of the surgical technique) is often difficult to determine until 3-4 years after the original surgery.

Management of the child with HD that is not doing well following their pull-through is complex and requires a systematic approach to exclude the common causes. (1) The likely reasons for poor functional outcomes include a twist in the colon, stricturing of the colo-anal anastomosis, an extrinsic muscular cuff in the pelvis, and/or residual aganglionosis at the distal end of the colon. Residual aganglionosis (or a transition zone pull-through) may be slow to present, difficult to diagnose, and problematic to treat. Ghosh *et al*. (2), in the description of their tertiary centre experience with transition zone pull-through, have highlighted the complexities inherent in management.

Residual aganglionosis is prevented during the initial pull-through procedure by the use of timely intra-operative frozen section pathology. The anatomical pathologist assesses the colonic specimen to determine the presence or absence of ganglion cells in the myenteric and submucosal plexi. (3) The expertise required for this

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assessment is limited to specially trained paediatric pathologists. Awareness of the inherent variability in distribution and quality of ganglia, as the colon transitions from aganglionic to ganglionic, has led many surgeons to change their intra-operative practice. Once the extent of aganglionosis has been confirmed, the surgeon will send a complete ring (doughnut) of tissue to confirm circumferential ganglionosis. Only once this is confirmed will the coloanal anastomosis be completed. However, in some tertiary centres where the required pathological expertise is lacking, the surgeon will perform the initial levelling biopsies at a separate operation to ensure the correct interpretation.

Ghosh *et al*. have described an incidence of transition zone pull-through of 16% that is comparable with other large published studies. Errors in histological interpretation were the major cause, with a significantly longer transition zone noted in affected patients. Of note, these patients also had more proximal extent of their disease, which may have led to more difficult pathological interpretation. Whilst none of the described patients has undergone re-do surgery for their residual aganglionosis, there is a high likelihood that they will require further interventions with increasing age. (4) Their risk of Hirschsprung-associated enterocolitis, a potentially fatal condition, is also likely to be increased. (5)

Finally, in addition to further refinement of surgical and pathological expertise, there remains the need for greater understanding of how the bowel transitions from normal

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to abnormal, beyond a simplistic description of presence or absence of ganglion cells.

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