IPO-05  Patchy innervation in pull-through bowel of Hirschsprung’s disease

Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine
Tsubasa Takahashi, Yoshifumi Kato, Tadaharu Okazaki, Hiroyuki Koga, Geoffrey Lane,
Atsuyuki Yamataka

Background: The effectiveness of pull-through for Hirschsprung’s disease (HD) is dependent on accurate identification of normally innervated bowel in intraoperative biopsy specimens, and conventionally, 2 cm above the level of normoganglionic biopsy sites has been regarded as being uniformly normoganglionic. We report a case of patchy innervation in a child with HD only identified by circumferential biopsying.

Case report: A 5-month-old boy was referred for investigation of HD. Caliber change was present in the sigmoid colon on contrast enema and rectal suction biopsy proved aganglionosis. During laparoscopy-assisted transanal endorectal pull-through, approximately 7×7×10 mm sized biopsies of bowel 2 cm proximal to the level of normoganglionosis confirmed by laparoscopic colon biopsy were performed at 3, 6, 9, and 12 o’clock for thoroughness. We were expecting all to be normoganglionic, but aganglionosis was found at 3 o’clock, suggestive of patchy innervation at this level. Circumferential biopsies were performed a further 2 cm proximally to exclude residual patchy innervation, and all sites were normoganglionic. This level was used for pull-through with excellent outcome.

Conclusion: Our case confirms that patchy innervation can be missed when conventional biopsies are normoganglionic. One way of preventing this would be to perform biopsies circumferentially, until ideally, all are normoganglionic. We recommend circumferential biopsies be performed routinely to prevent bowel with patchy innervation from being used for pull-through and possibly causing postoperative bowel dysmotility in a subgroup of HD patients.