IS-1

Altered distribution of interstitial cells of Cajal in motility disorders

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Introduction

Interstitial cells of Cajal (ICCs) are specialised pacemaker cells in the gastrointestinal tract that are of mesenchymal origin and fundamental for the physiological functions of the gastrointestinal smooth muscles. ICCs facilitate active propagation of electrical events and mediate neurotransmission. ICCs are present as myenteric ICCs (ICCs-MY) or intramuscular ICCs (ICCs-IM).

The aim of this study was to investigate the distribution of ICCs in the normal bowel and bowel of Hirschprung's disease (HD), total colonic aganglionosis (TCA), isolated hypoganglionosis (IH), intestinal neuronal dysplasia (IND), megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS), and internal anal sphincter achalasia (IASA).

Material and Methods

Full thickness bowel biopsies were obtained from patients with: HD (n=10), TCA (n=4), IH (n=6), IND (n=6), MMIHS (n=4), IASA (n=8). Normal control specimens of large bowel and internal anal sphincter were obtained from children without motility disorders. Single immunohistochemistry staining using monoclonal and polyclonal anti c-kit antibodies for frozen or paraffin sections. Double staining NADPH-D histochemistry/c-kit immunohistochemistry in whole-mount preparation were carried out. Sections were examined in light and confocal scanning microscopy.

Results

There was lack or marked reduction of ICCs-My and ICCs-IM in HD, TCA, IH, IND, and IASA specimens compared to controls. In the MMIHS samples only ICCs-IM were reduced whereas the distribution of ICCs-MY was similar to the normal bowel.

Conclusion

Altered distribution of ICCs in the gut may interfere with neurotransmission and normal propagation of electrical events, causing motility dysfunction in HD, TCA, IH, IND, MMIHS and IASA patients.