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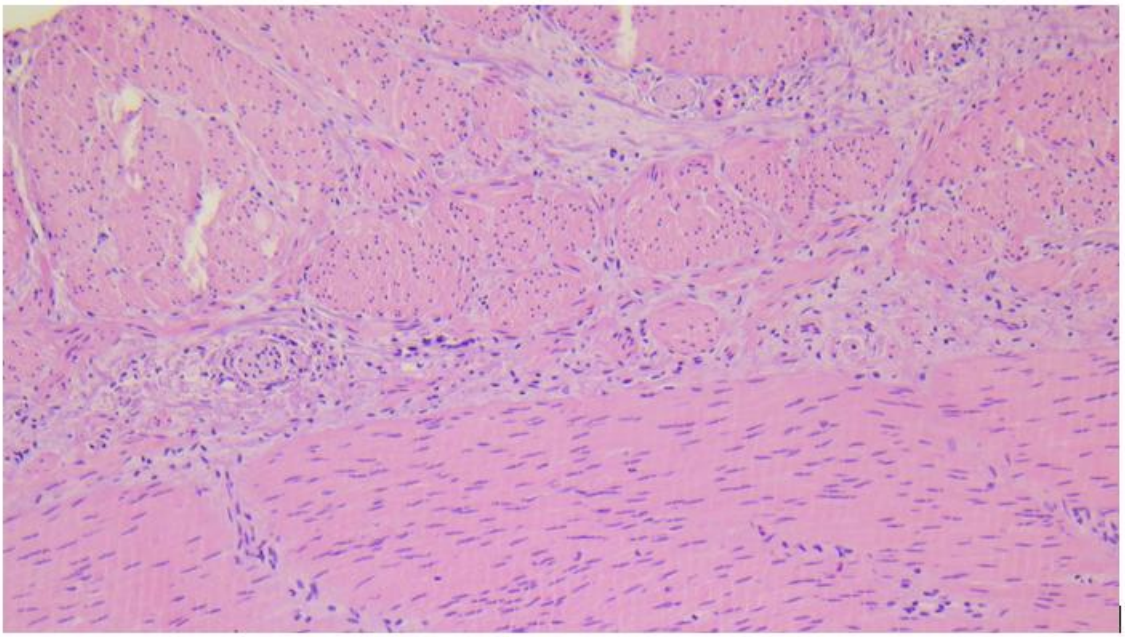
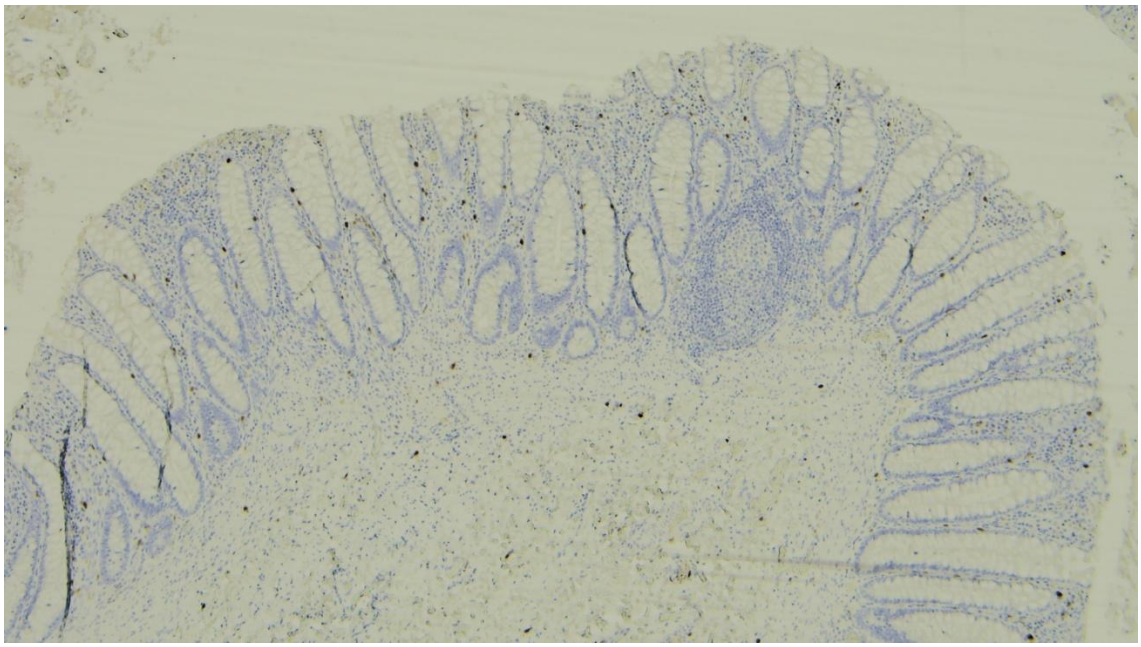
Patient 1, affected with HRSC disease



Supplementary Figure 1: Histological image (hematoxylin-eosin). Histological examination showed an aganglionic colon tract with longitudinal and circular muscular layers**.**



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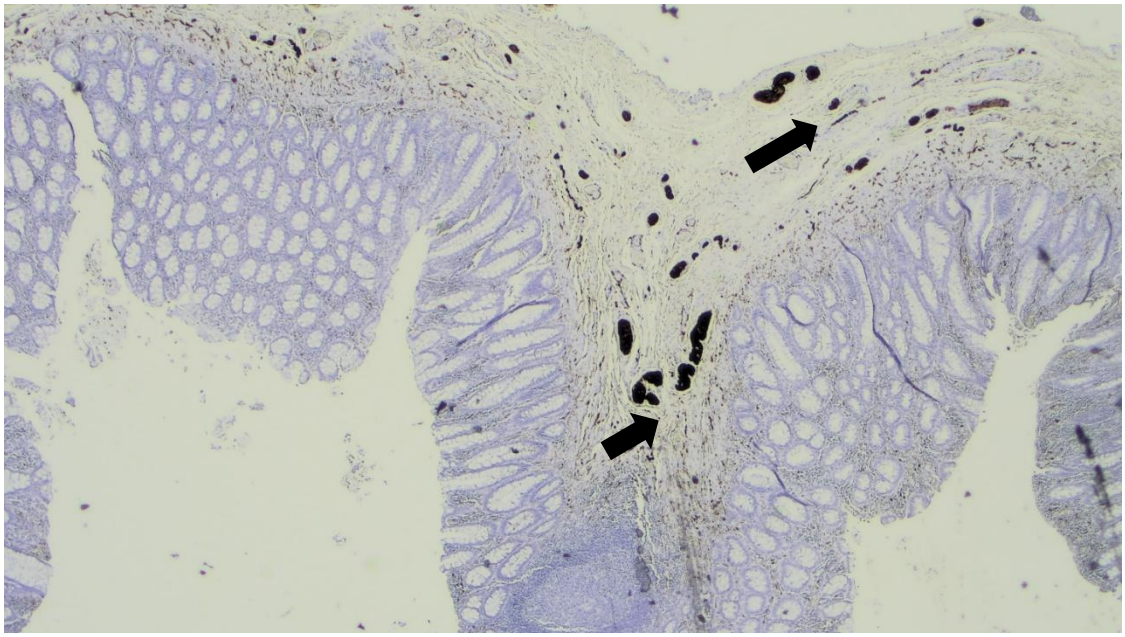
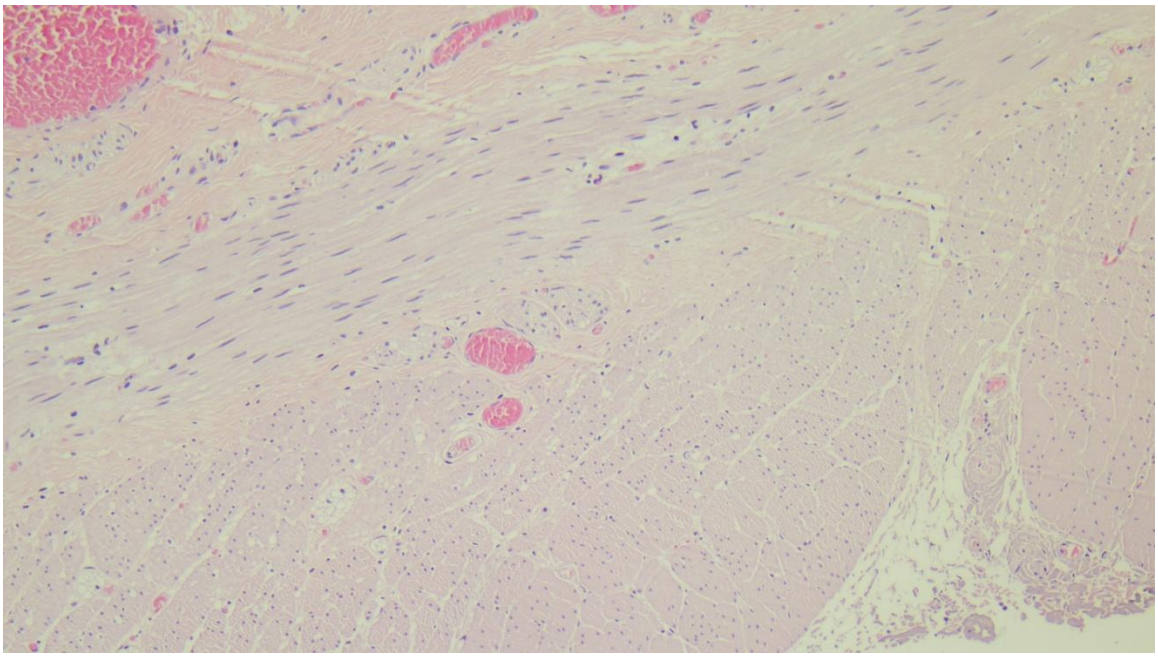
Supplementary Figure 2: Histological image (calretinin). Immunohistochemical investigation for Calretinin showed no submucosal ganglia and no neurites in the lamina propria. Scattered positive mast cells were present, useful as an internal control.



Supplementary Figure 3: Histological image (S100). Immunohistochemical investigation for S100 showed several hypertrophic nerves in the submucosa (arrows).

Patient 2, affected with HRSC disease



Supplementary Figure 4: Histological image (hematoxylin-eosin). Histological examination showed absence of submucosal and myenteric ganglia and presence of hypertrophic nerves in the wall.

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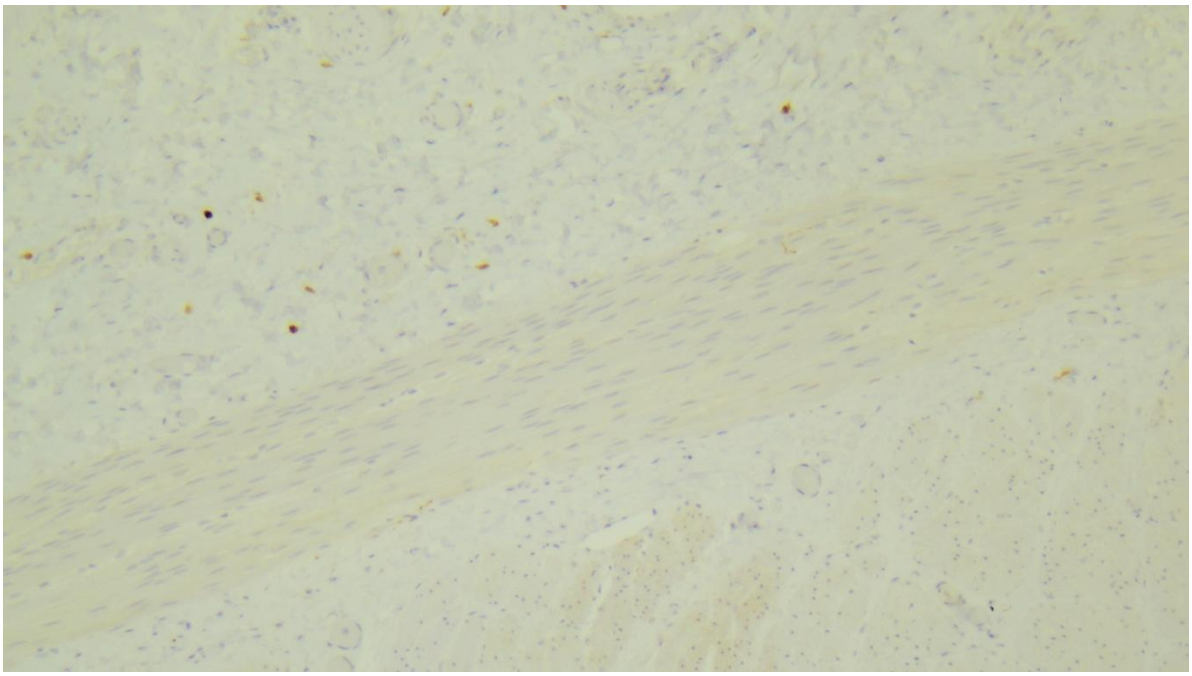
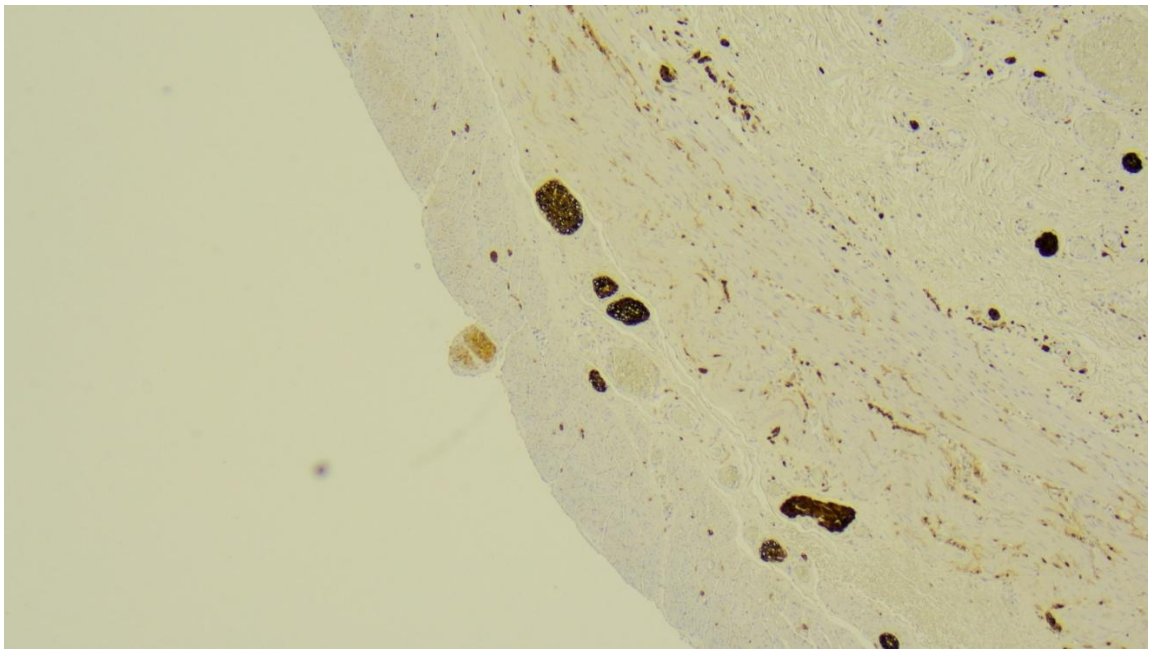


Supplementary Figure 5: Histological image (calretinin). The immunohistochemical investigation Calretinin confirmed the absence of ganglion structures and absence of innervation of the lamina propria. Rare and scattered positive mast cells were present in the image.



Supplementary Figure 6: Histological image (S100). Immunohistochemical investigation with S100 showed several hypertrophic nerves in the muscularis propria and submucosa.

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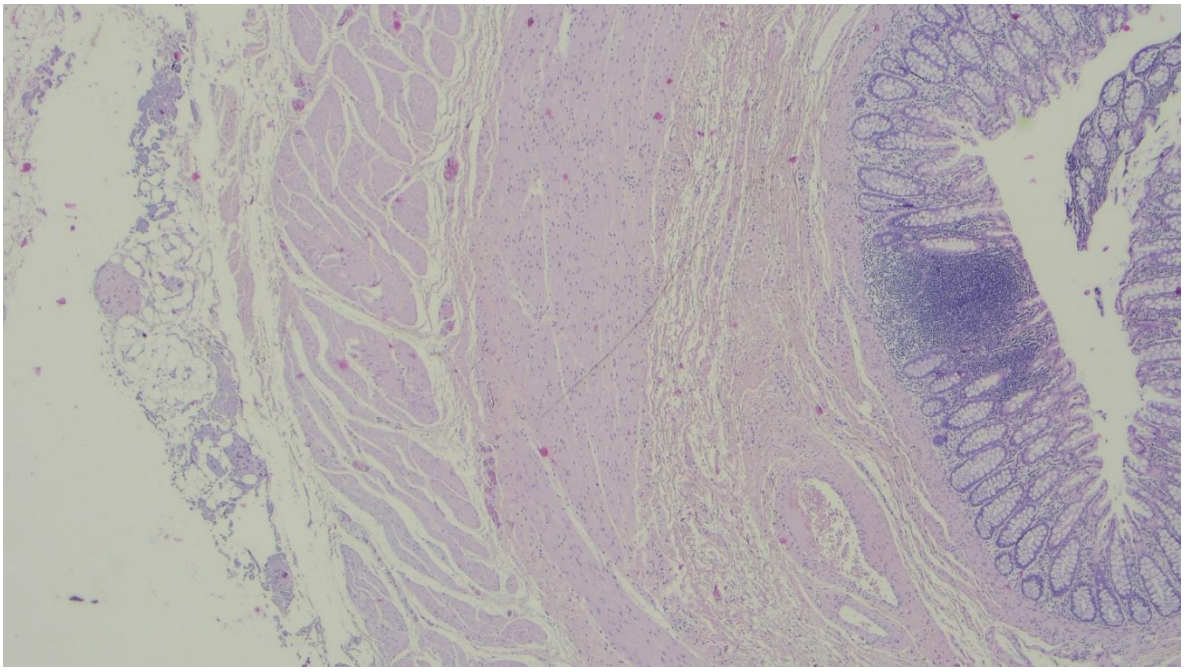
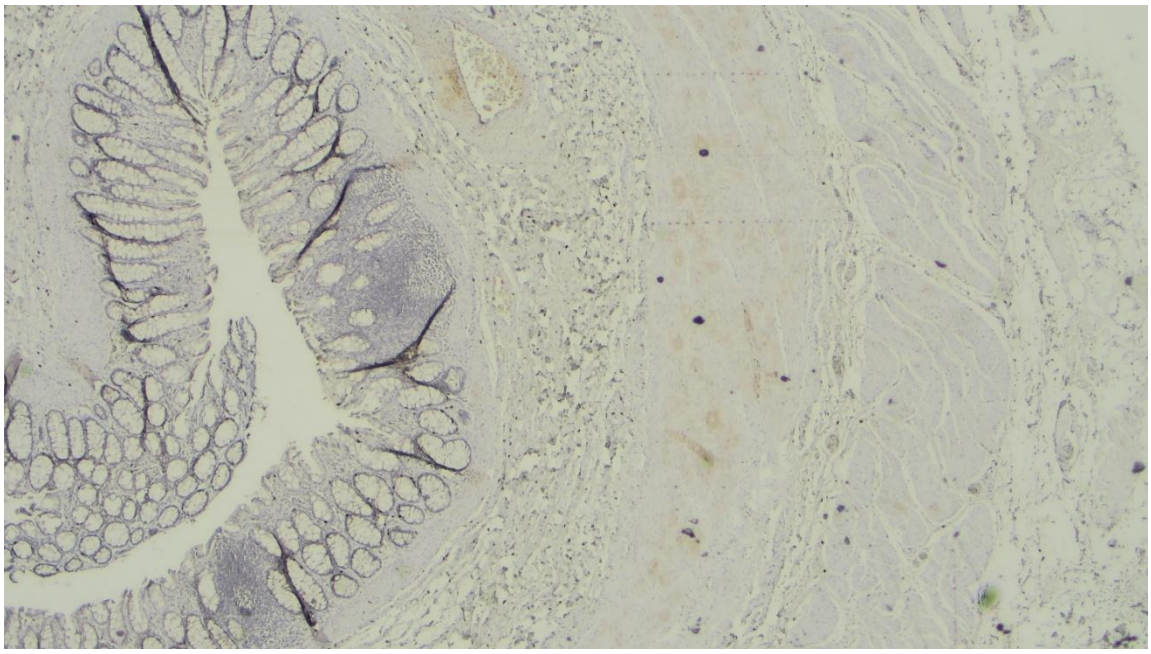
Patient 3, affected with HRSC disease



Supplementary Figure 7: Histological image (hematoxylin-eosin). Histological examination showed a thickened colic wall totally devoid of ganglia in the submucosal and myenteric plexuses.



Supplementary Figure 8: Histological image (calretinin). Calretinin immunohistochemical investigation confirmed the absence of ganglia and the absence of lamina propria innervation.

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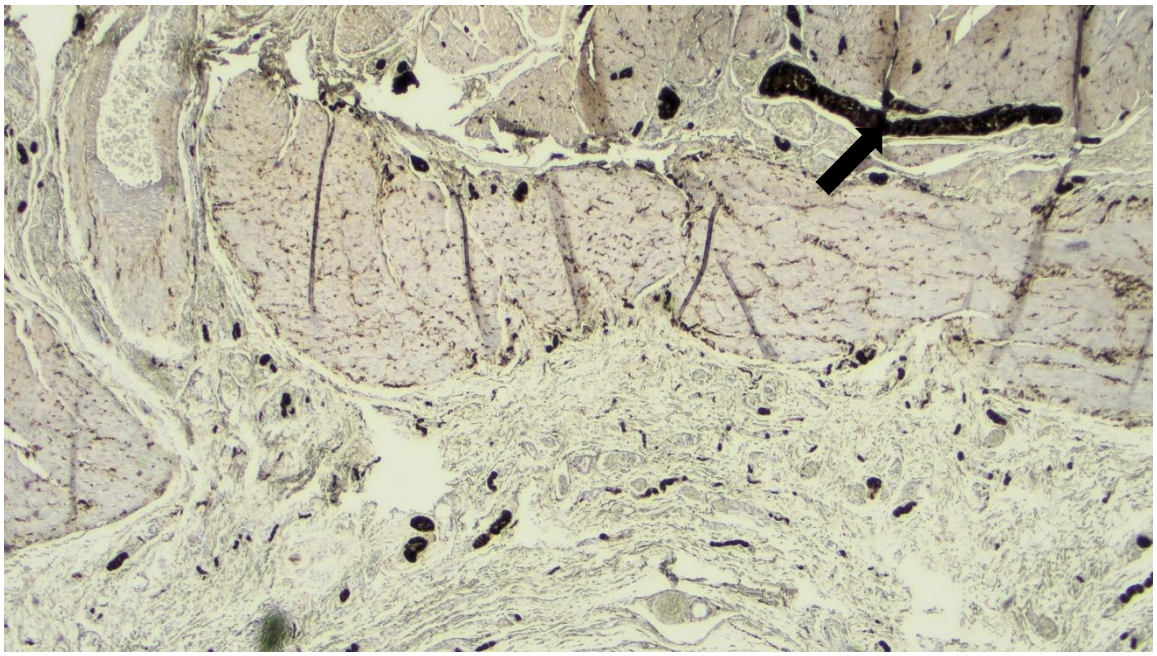
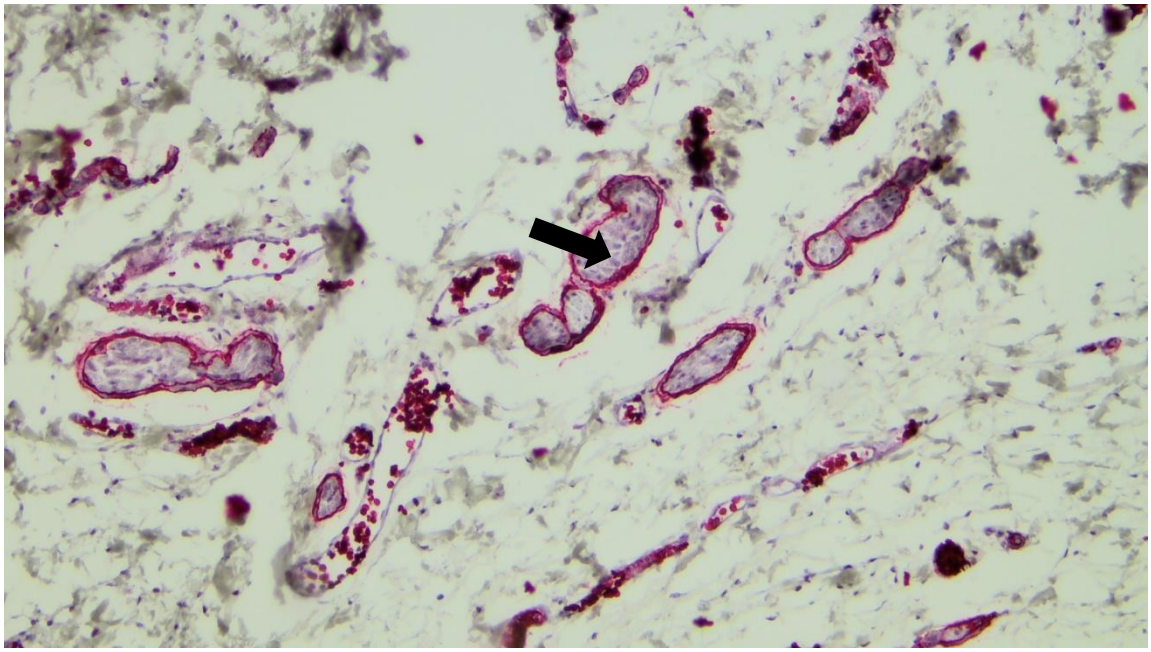


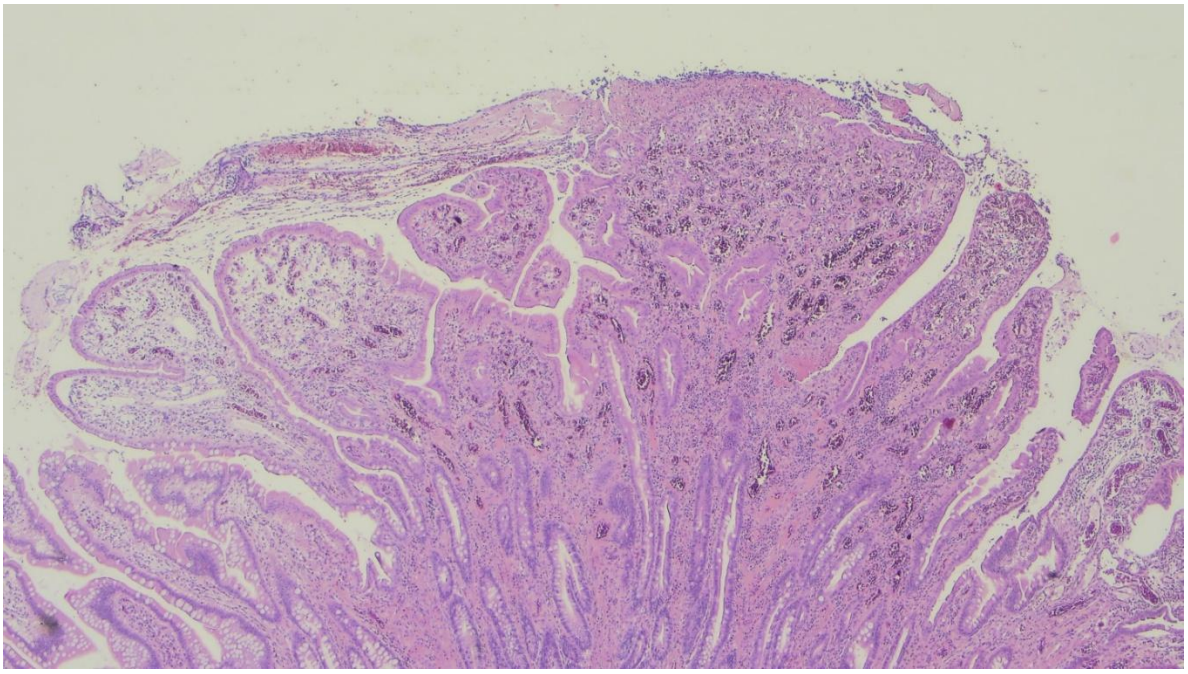
Supplementary Figure 9: Histological image (S100). The S100 immunohistochemical investigation showed numerous hypertrophic nerves in the intestinal wall (arrow).



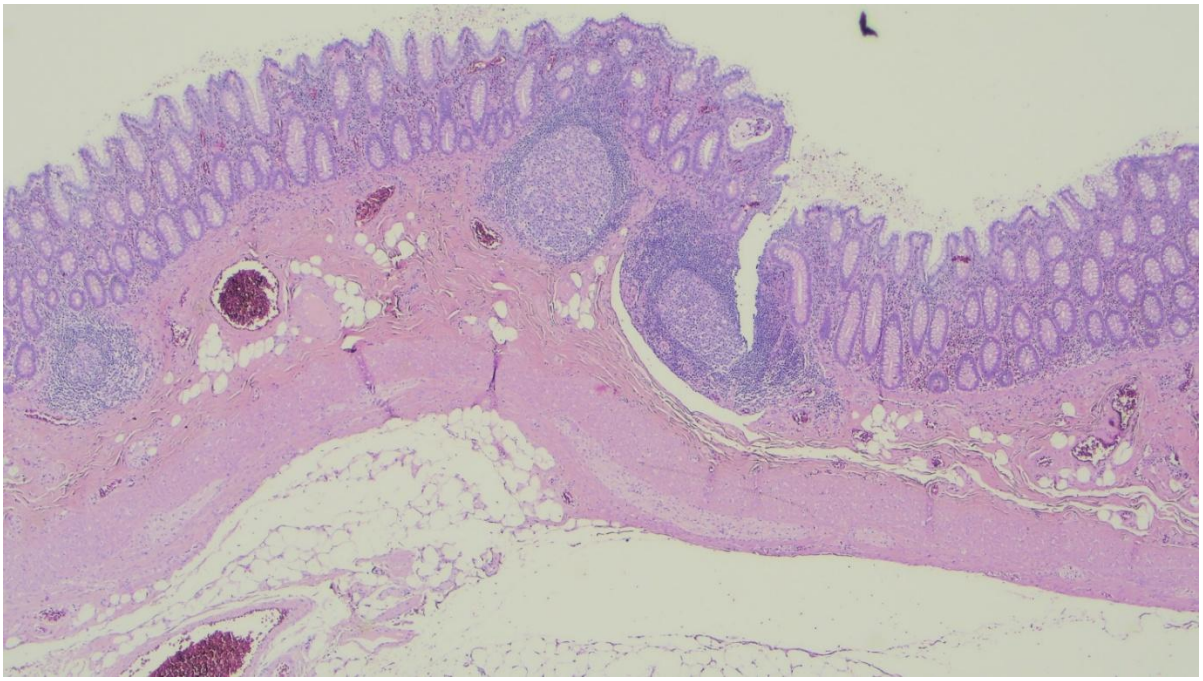
Supplementary Figure 10. Histological image (GLUT1). GLUT1 immunohistochemical investigation showed

hypertrophic nerves with perineurium (arrow).

Patient 4, affected with myogenic PIPO



Supplementary Figure 11: Histological image (hematoxylin-eosin). The ileal mucosa showed numerous erosions and inflammatory polyps covered with granulation tissue.



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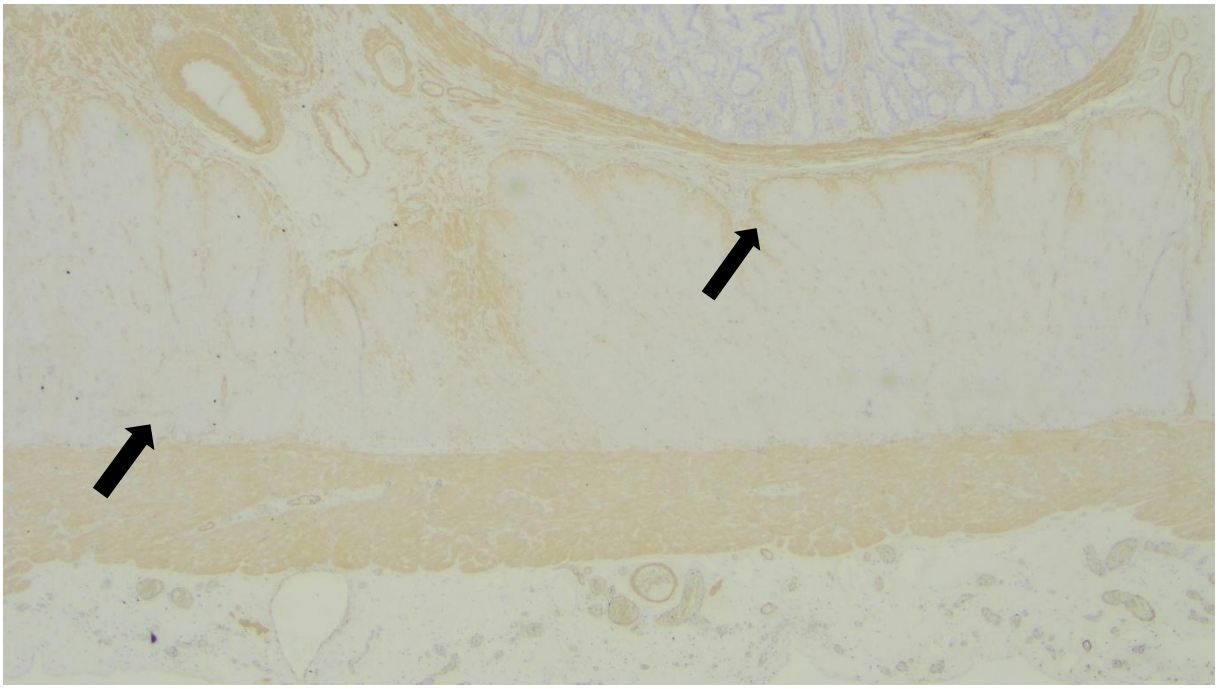
Supplementary Figure 12: Histological image (hematoxylin-eosin). The colic wall showed marked thinning of the muscle wall.



Supplementary Figure 13: Histological image (α-SMA). Immunohistochemistry investigation for smooth muscle actin showed abnormal expression of the antibody: loss of expression by the inner circular layer with preserved expression of a thin rim of internal muscle (arrows).

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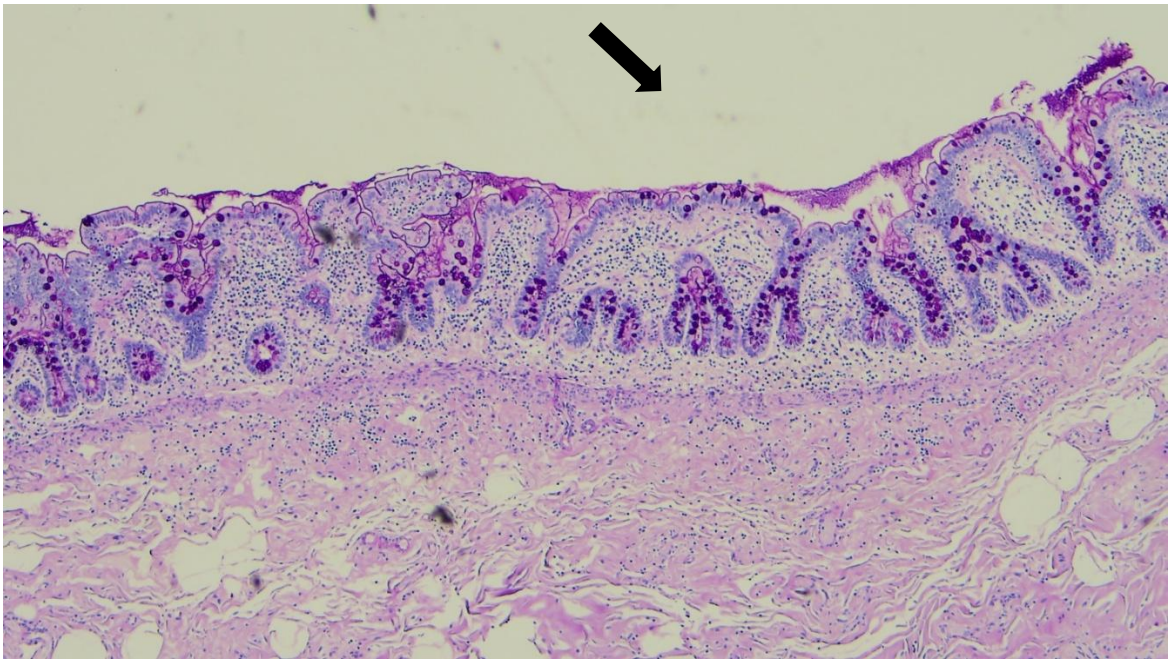
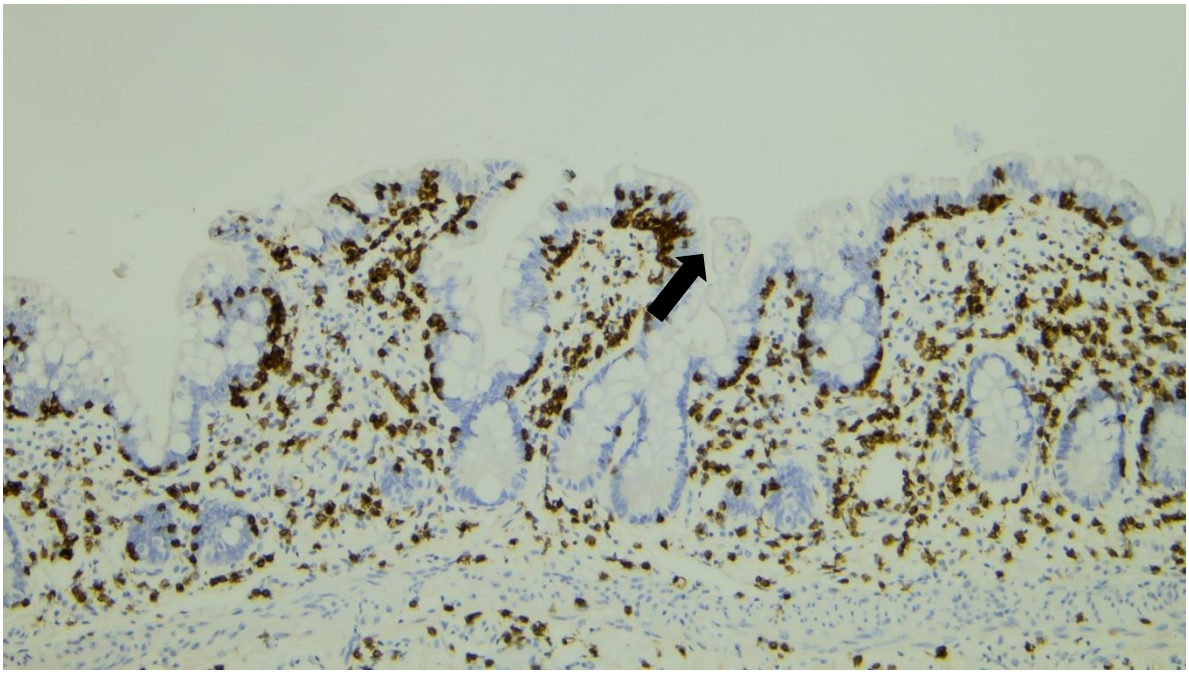
Patient 6, affected with mitochondrial PIPO



Supplementary Figure 14: Histological image (hematoxylin-eosin). The ileal mucosa showed villous atrophy (arrow). PAS-D histochemical staining.



Supplementary Figure 15: Histological image (CD3). CD3 immunohistochemistry investigation showed intraepithelial lymphocytosis (arrow).