Endoscopic management of obstructive pancreatitis with a metal stent in two family members with hereditary cationic trypsinogen (*PRSS1*) deficiency

Fig. 1 Fluoroscopic view of the pancreatic duct of a 37-year-old woman with genetically proven hereditary cationic trypsinogen (*PRSS1*) deficiency pancreatitis: **a** before endoscopic treatment; **b** after endoscopic treatment.

Fig. 2 Fluoroscopic view of the pancreatic duct of the 12-year-old daughter of the patient of **Fig. 1**: **a** before endoscopic treatment; **b** during endoscopic treatment.

We report here the case of a 37-year-old woman and her daughter (now 12 years old) with genetically proven hereditary cationic trypsinogen (*PRSS1*) deficiency pancreatitis. The two patients were treated in a similar manner: endoscopic extraction of large obstructive stones located mainly in the head of the pancreas upstream of a stricture of the distal (cephalic) pancreatic duct and associated with extreme dilation (10–15 mm) of the pancreatic duct, manifesting as bouts of mild acute pancreatitis and abdominal pain that could not be relieved with analgesics (**Fig. 1–3**).

Both patients underwent pancreatic sphincterotomy, hydrostatic dilation of

the pancreatic duct stricture with a 6-mm Hurricane RX Biliary Dilation Balloon (Boston Scientific, Natick, Massachusetts, USA), removal of the stone fragments with a Dormia basket, and placement of an 8-mm-diameter, 60-mm-long fully covered self-expandable metal stent (SEMS) (WallFlex Biliary RX Stent; Boston Scientific) to dilate the stricture and facilitate removal of the remaining stone fragments (**Fig. 4** and **Fig. 5**). The mother's stent was left in place for 1 month and the daughter's for 2 weeks. After stent removal, the mother experienced one episode of abdominal pain, with no further symptomatic relapse so far. Because of two flares of recurrent

mild pancreatitis, the daughter underwent additional endoscopic therapeutic sessions; these consisted of a second stricture dilation, followed by placement of another fully covered SEMS for 3 months (same device) and subsequent placement of a 10-Fr plastic stent (Johlin Stent; Cook Medical, Winston-Salem, North Carolina, USA), which was withdrawn 3 months later. Since this procedure, the daughter has remained asymptomatic and has resumed school attendance uneventfully. Optimal stenting modalities have not been detailed in the rare reports devoted to patients with this condition [1–5]. The use of fully covered SEMS was of particular interest in these two patients, who presented with similar findings (short distal stricture and unusually large upstream dilation), because these stents can facilitate the removal of large stones and reduce the discrepancy between duct size in the head and body of the pancreas, a major factor in stone formation.

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Competing interests: None

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Fig. 3 Endoscopic ultrasound views of the patient in [Fig. 2](#) (the daughter): **a, b** stones in the dilated pancreatic duct; **c** after treatment, stones in the pancreatic parenchyma (whereas the main pancreatic duct is free of stones); **d** the pancreatic duct after treatment, exhibiting a moderately enlarged caliber.

Fig. 5 Duodenoscopic view of the removal of stone fragments with the Dormia basket.

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Fig. 4 Fully covered self-expandable metal stent in the pancreatic duct.