Recurrent Biliary Obstruction Overcome with Metal Biliary Stent Placement in the Surgically Altered Down Syndrome Patient for Congenital Duodenal Atresia

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Purpose

- Patients with Down syndrome have increased rates of GI malformations and pathology
- In our case a patient with Down syndrome had surgical interventions at birth for congenital duodenal atresia and recurrent cholelithiasis
- Duodenal atresia is a failure of the duodenum to canalize in gestation resulting in gastric outlet obstruction at birth, and it is largely associated with Down syndrome
- It requires early surgical duodenoduodenostomy or duodenojejunoanastomosis
- Patients with Down syndrome also have a high prevalence of cholelithiasis which may require surgery
- Historically, surgical procedures like cholecdochoduodenostomy were used in patients with variants of biliary lithiasis
- We describe a unique endoscopic intervention in an adult patient with Down syndrome with recurrent biliary obstruction and conjoined anatomy after childhood surgeries

Case Presentation

- A 53 year-old female with remote cholecdochoduodenostomy and duodenojejunostomy presents with recurrent cholelithiasis
- The patient has a history of Down syndrome with duodenal atresia requiring surgery at birth, and cholecystectomy later in life
- She had recurrent episodes of cholelithiasis requiring percutaneous transhepatic cholangiography by interventional radiology (IR)
  - Interventions included balloon dilation of the ampulla and internal/external biliary tube placement
  - She still continued developing stones at the cholecdochoduodenostomy anastomotic site with abdominal pain and elevated transaminases
  - Her recurrent tube malposition and stone formation led to four IR procedures for tube repositioning and stone removal
  - Endoscopic intervention was necessitated by failed IR therapies and patient discomfort with the external tube
  - A rendezvous procedure with IR was initially performed to define positioning
    - The standard duodenoscope could not reach the anastomotic site and was exchanged for a pediatric colonoscope
    - Antegrade cannulation with a wire via transhepatic access allowed retrograde guidance of the colonoscope to the anastomotic site
  - A subsequent ERCP was performed using a pediatric colonoscope based on previously defined anatomy
    - Cholangiogram, balloon sweeps with stone removal, and fully covered biliary metal stent deployment were performed
  - In this case deviation from the standard duodenoscope used in ERCP facilitated the definitive therapy in a patient with prior choledochoduodenostomy
  - At follow-up the patient is doing well and her transaminases have normalized

Discussion

- To our knowledge this is the first case of an endoscopic intervention in altered surgical anatomy for duodenal atresia in a patient with Down syndrome
- Gastroenterologists should become familiar with congenital GI abnormalities with potential surgical alterations to prepare for interventions in these patients

References:

Lehigh Valley Health Network, Allentown, PA

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