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

Patrick Hickey DO
Lehigh Valley Health Network, Patrick.Hickey@lvhn.org

Matthew Sullivan DO
Lehigh Valley Health Network, Matthew.Sullivan@lvhn.org

Saba Ahmad MD
Lehigh Valley Health Network, Saba.Ahmad@lvhn.org

Hiral Shah MD
Lehigh Valley Health Network, hiral_n.shah@lvhn.org

Published In/Presented At

Hickey, P., Sullivan, M., Ahmad, S. & Shah, H. (2014, October, 17). *Recurrent Biliary Obstruction Overcome with Metal Biliary Stent Placement in the Surgically Altered Down Syndrome Patient for Congenital Duodenal Atresia.* Poster session presented at the American College of Gastroenterology Annual Scientific Meeting, Philadelphia, PA.

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

Patrick Hickey DO, Matthew Sullivan DO, Saba Ahmad MD, Hiral Shah MD

Lehigh Valley Health Network, Allentown, PA

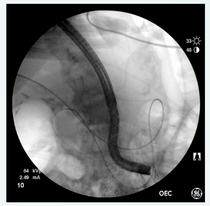
Purpose

- 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 recanalize in gestation resulting in gastric outlet obstruction at birth, and it is largely associated with Down syndrome
- It requires early surgical duodenoduodenostomy or duodenojejunostomy
- Patients with Down syndrome also have a high prevalence of cholelithiasis which may require surgery
- Historically, surgical procedures like choledochoduodenostomy 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 convoluted anatomy after childhood surgeries

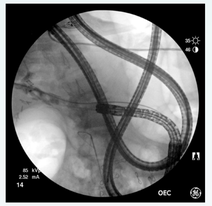
Case Presentation

- A 53 year-old female with remote choledochoduodenostomy and duodenojejunostomy presents with recurrent choledocholithiasis
- 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 choledocholithiasis 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 choledochoduodenostomy 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

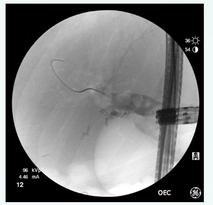
Images



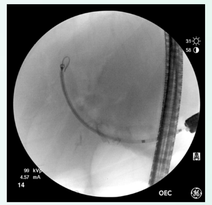
Transcutaneous catheter with a wire traversing internally through biliary anastamosis into the efferent limb, and a pediatric colonoscope is seen identifying the wire at the enteral anastomosis



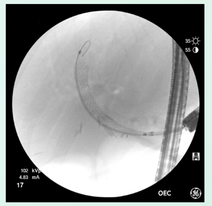
A pediatric colonoscope directly visualizing the biliary enteric anastomosis after localization with transcutaneous wire guidance



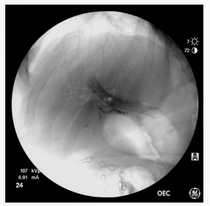
Cholangiogram showing significant dilatation of the bile duct (12 mm in cross-sectional diameter) with multiple stones appreciated in the distal common bile duct



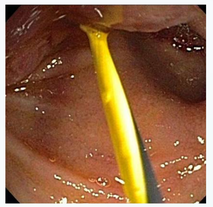
A fully covered self-expanding metal stent is directed into the biliary anastomosis



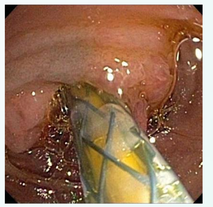
A partially deployed biliary stent at the site of biliary anastomosis



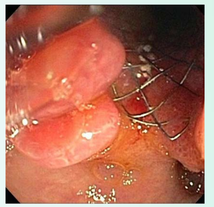
Well positioned biliary stent at the biliary anastomosis placed using a pediatric colonoscope



Wire guided cannulation of the choledochoduodenostomy site using a pediatric colonoscope



Placement of a biliary stent at the anastomotic site using a pediatric colonoscope



Biliary metal stent after deployment at the biliary anastomosis

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:

1 Demirel B, Kekilli M, Sahin B, et al. ERCP experience in patients with choledochoduodenostomy: diagnostic findings and therapeutic management. *Surgical Endoscopy.* April 2011;25(4):1043-1047.

2 Yamaner S, Bilsel Y, Sokucu N, et al. Endoscopic diagnosis and management of complications following surgery for gallstones. *Surgical Endoscopy* [serial online]. December 2002;16(12):1685-1690.