A Case of Congenital Duodenal Web Causing Duodenal Stenosis in a Down Syndrome Child: Endoscopic Resection with an Insulated-Tip Knife

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A 35-month-old girl visited our hospital with repetitive vomiting and abdominal distention; this was especially aggravated after the introduction of solid and semisolid foods. At 5 months of age, the patient, who had Down's syndrome, had undergone surgery for ventricular septal defect, atrial septal defect, and patent ductus arteriosus, and had subsequently been frequently hospitalized for respiratory infections and other viral infectious diseases. After her admission, the abdominal distension improved with fasting and intravenous fluid therapy. Radiograph from a small-bowel series revealed a thin filling defect with a dilated duodenal bulb in the distal region of the second portion of the duodenum, suggesting a duodenal web, and endoscopy revealed duodenal stenosis. We therefore performed endoscopic resection with an insulated-tip knife because of the history of prior operations, fasting problems after operations, and respiratory infections. Seven days later, scar formation was noted on the second portion of the duodenum, the scope passed well at the excision site, and no retained food material was noted on the follow-up endoscopy. After the procedure, the patient's abdominal distention and repetitive vomiting subsided, and she was discharged with the ability to eat an age-appropriate normal diet. There were no specific symptoms or other complications for 1 year after the procedure. (*Gut Liver* 2011;5:105-109)

Key Words: Insulated-tip knife; Endoscopic electrocauterization; Duodenal stenosis; Duodenal web; Down syndrome

INTRODUCTION

The association between Down syndrome and gastrointestinal abnormalities is well known. Down syndrome is recognized as a possible predisposing condition for gut abnormalities and

congenital heart disease as part of the VATER syndrome.¹ Patients with congenital malformations of the duodenum, such as duodenal stenosis, atresia, and annular pancreas, develop early signs and symptoms in the neonatal period and belatedly in late childhood. There are some intra-operation and post-operation difficulties when performing general anesthesia in children who show symptoms of Down syndrome and intestinal stenosis and who need to have surgery for gut disease.¹ Considering the multiple problems related to their underlying congenital heart disease, frequent respiratory infections, and other possible abnormalities, a conventional laparotomy or a laparoscopic duodenoduodenostomy for treatment may be invasive and risky. Indeed, this may not be a good choice for children with Down syndrome, especially for those who have scars on the chest wall, indicating that they have had a previous operation for congenital heart disease. There are many reports about endoscopic treatments with an insulated-tip (IT) knife in adults who have an acquired stenosis, and this is regarded as a favorable and effective procedure. There are very few reports about the IT-knife in children, however.²⁻⁴ We report a case of a duodenal web in a Down syndrome child that caused duodenal stenosis, in which the duodenal web was directly excised with an IT-knife without complications.

CASE REPORT

A 32-month-old girl came to Gachon University Gil Medical Center because of acute, severe repetitive vomiting and abdominal distention. At 5 months, she was diagnosed with Down syndrome and had surgery for correction of a ventricular septal defect, an atrial septal defect, and a patent ductus arteriosus. Afterward, she was often admitted and treated for respiratory infections and other viral infectious diseases. She had a history since infancy of frequent constipation and occasional postpran-

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Received on November 10, 2009. Accepted on April 6, 2010.

pISSN 1976-2283 eISSN 2005-1212 DOI: 10.5009/gnl.2011.5.1.105

Fig. 1. The findings of the gastrointestinal barium study. (A) A beak-like narrowing of the distal esophagus up to the gastroesophageal junction with mild passage disturbance, and a thin filling defect are noted on the distal region of the second portion of the duodenum. (B) Contrast is retained on the dilated stomach and duodenal bulb.

dial vomiting, and the postprandial vomiting became aggravated with the introduction of semisolid or solid foods. Two days prior to admission, she had repetitively vomited bilious food mixed with undigested particles over 15 times per day. Oral intake became intolerable, and she was admitted for fluid therapy. On the day of admission, her body temperature was 36.8°C with a pulse rate of 126 beats/min, a respiratory rate of 24 breaths/min, and a blood pressure of 90/50 mm Hg. Her body weight was 10.6 kg, and her height was 84 cm. Physical examination revealed a Down facies and slightly coarse breath sounds, and regular heart beats were heard from a chest with scars from previous operations. Her abdomen was moderately distended without tenderness or rebound tenderness. Results of laboratory studies were as follows: white blood cell count 9,160/mm³, platelet count 399,000/mm³, and hemoglobin 12.7 g/dL. The results of tests were in normal ranges. A plain abdominal radiograph showed a mild dilatation of the duodenum but no definite double bubble sign. The upper GI series showed a beak-like narrowing of the distal esophagus up to the gastroesophageal junction with mild passage disturbance and a thin filling defect with a dilated duodenal bulb in the distal region of the second portion of the duodenum (Fig. 1). On a small bowel series, contrast showed a thin filling defect with a dilated duodenal bulb in the distal region of the second portion of the duodenum, suggesting a duodenal web (Fig. 2). A flexible endoscope was then inserted into the duodenum and revealed a membranous stenosis in the second portion of the duodenum, suggesting a duodenal web (Fig. 3). After fasting and fluid treatment, her abdominal distention and vomiting slightly improved. But she needed ultimate treatment for the symptomatic duodenal web. Conventional laparotomy or a laparoscopic duodenoduodenostomy were considered, but given her underlying condition and past medical history, endoscopic electrocauterization was instead attempted to remove the mucosal diaphragm of the stenotic lumen caused by the duodenal web. Avoiding the Papilla of Vater, the diaphragm of the duodenal web was cautiously resected and electrocauterized with an IT-knife (Fig. 4). After the procedure, she had an uneventful course without any bleeding, perforation or

Fig. 2. Findings of the small-bowel series. A thin filling defect with a dilated duodenal bulb is noted in the distal region of the second portion of the duodenum.

other complications. She had restarted oral feeding 1 day after the endoscopic procedure, and she underwent a follow-up endoscopy 7 days after the endoscopic resection. On the follow-up endoscopy, scar formation was seen on the second portion of the duodenum at the site of cauterization, and no passage disturbance through the duodenum or retained food material were noted (Fig. 5). After that, the patient's abdominal distention and repetitive vomiting improved, and she was discharged with the ability to eat an age-appropriate normal diet. She has been free from any specific symptoms for 14 months after the endoscopic procedure.

DISCUSSION

Stenosis and atresia are common birth defects that affect the small intestine, and they can affect multiple sites of the intestine. The most frequently affected site is the duodenum, while the ileum is the least affected.⁵ The incidence of duodenal stenosis and atresia has been reported to be approximately 2-5 per 10,000 live births.⁵ Associated congenital anomalies have been reported in more than 50% of affected patients and can include

Fig. 3. Initial endoscopic findings. (A) A mucosal diaphragm is seen on the second portion of the duodenum, suggesting a duodenal web. (B) A mucosal diaphragm in the second portion of the duodenum shows a stenotic lumen.

Fig. 4. Endoscopic findings. (A) The mucosal diaphragm of a stenotic lumen caused by a duodenal web was resected and electrocauterized with an insulated-tip (IT) knife. (B) A stenotic lumen is dilated after the removal of the diaphragm with an IT-knife.

Fig. 5. The follow-up endoscopic findings. (A) Scar formation is noted on the second portion of the duodenum at the site of cauterization. (B) The lumen is shown in the second portion of the duodenum with no passage disturbance.

congenital heart disease, pancreatic anomalies, intestinal malrotation, esophageal atresia, variants of imperforate anus, renal anomalies or other combinations of anomalies such as VATER/VACTERALS.^{1,6,7} Chromosomal abnormalities, particularly Down syndrome, occur in approximately 15-27% of affected patients.⁵ Patients with duodenal stenosis or atresia usually develop obstructive symptoms in infancy, and the initial diagnosis is generally made in the neonatal period or at least by late childhood.⁸

The clinical presentation of affected patients includes intermittent, recurrent bilious vomiting, upper abdominal distension, and sometimes postprandial right upper abdominal pain, but the symptoms may be ignored because of the associated retardation of children with Down syndrome.^{9,10}

The primary diagnosis is made by abdominal radiographs, and a plain abdominal radiographs shows dilatation of the duodenum and disturbed transit of contrast media with a filling

defect in the second portion of the duodenum in an infant or child with recurrent protracted vomiting.¹¹ Flexible endoscopy can reveal the overextension of the duodenum, the protrusion of the duodenal web and, if any, the mucosal diaphragm in the stenotic lumen of the duodenum.¹¹⁻¹³ Duodenoduodenostomy, duodenostomy, duodenotomy with incision or excision of the web or diaphragm, duodenojejunostomy, or duodenoplasty can all be considered as different modes of operative management. The treatment of choice for duodenal stenosis and atresia is duodenoduodenostomy, and exploratory laparotomy is necessary for a second web at the time of operation.^{1,11} A child with Down syndrome often needs multiple operations because of associated anomalies, such as cardiac defects, anorectal defects or tracheoesophageal defects.^{11,14} Operative techniques and systems of care have recently improved, although the possibility of complications and mortality rates after multiple operations still remain high.^{1,11,15} As a result, a higher risk of intra- and post-operative complications and a higher long-term mortality rate have been reported in children who have intestinal stenosis combined with Down syndrome.^{1,15} The patient with duodenal stenosis must undergo surgical correction of the obstruction, and the suspected diagnosis will be confirmed at laparotomy. However, considering the multiple problems related to underlying congenital heart disease, frequent respiratory infections, and preceding multiple operations, a conventional laparotomy or a laparoscopic duodenoduodenostomy for treatment of a Down syndrome patient with intestinal stenosis may be invasive and risky.^{16,17} Furthermore, consideration of the cosmetic aspect must be given to children with previous scars. Recently, endoscopic electrocauterization has been performed in older children and adults with acquired stenotic problems and is regarded as a more favorable and effective procedure.¹⁸⁻²¹ Even though endoscopic mucosal resection has been established as one of the treatment options for early gastric cancer patients who have no risk of lymph node metastasis and is a good treatment of choice for other stenotic conditions, the most significant limitation of conventional endoscopic resection is the size of the resected specimen, that is, the difficulty in en bloc resection.²²⁻²⁴ The technique of using an IT-knife has been applied to endoscopic therapy, and endoscopic membranectomy and submucosal dissection over an extensive mucosal area are possible and have been conducted uneventfully.^{3,4,18,19} Particularly, a diaphragm type of duodenal web can be safely resected as much as possible by endoscopic electrocauterization with an IT-knife because it consists of mucosa and submucosa.^{20,25} We report here on a case of duodenal web causing duodenal stenosis in a Down syndrome child, and the duodenal web was directly resected with an IT-knife without complications.

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