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

Sang Seon Lee*, Seon Tae Hwang*, Nam Gil Jang†, Hann Tchah*, Duk Young Choi‡, Hyun Young Kim§, and Eell Ryoo*

Departments of *Pediatrics, ‡Emergency Medicine, †Pediatric Cardiology, and §Surgery, Gachon University of Medicine and Science Graduate School of Medicine, Incheon, Korea

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 distention 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. 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. 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. 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-
dial vomiting, and the postprandial vomiting became aggravat-
ed 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 in-
take 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 labora-
tory 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 radi-
ograph showed a mild dilatation of the duodenum but no definite
double bubble sign. The upper GI series showed a beak-like nar-
rowing of the distal esophagus up to the gastroesophageal junc-
tion with mild passage disturbance and a thin filling defect with
a dilated duodenal bulb in the distal region of the second por-
tion 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 disten-
tion and vomiting slightly improved. But she needed ultimate
treatment for the symptomatic duodenal web. Conventional
laparotomy or a laparoscopic duodenoduodenostomy were con-
sidered, 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 dia-
aphragm of the duodenal web was cautiously resected and elec-
trocauterized with an IT-knife (Fig. 4). After the procedure, she
had an uneventful course without any bleeding, perforation or
other complications. She had restarted oral feeding 1 day after
the endoscopic procedure, and she underwent a follow-up en-
doscopy 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 dis-
turbance 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 intest-
ate. The most frequently affected site is the duodenum, while
the ileum is the least affected. The incidence of duodenal ste-
nosis and atresia has been reported to be approximately 2-5 per
10,000 live births. Associated congenital anomalies have been
reported in more than 50% of affected patients and can include