Anal Canal Duplication in Infants

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Background/Purpose: Anal canal duplication (ACD) is the most distal and the least frequent digestive duplication. A review of the English-language literature found 15 cases reported in the pediatric age group.

Methods: A retrospective chart review was performed for our experience from 1999 to 2001 with 6 patients who presented with a midline postanal opening.

Results: All of 6 patients were girls and included one set of twins. Patients were from 3 to 9 months of age at the time of diagnosis (mean, 4.5 months). The anal canal duplication was delineated clearly by contrast study of the tract. All were tubular structures, 10 to 12 mm in length. None of the patients had any other associated anomalies. Five of 6 patients underwent operation between the ages of 3 and 8 months (mean, 5.4 months). Excision of the ACD was accomplished through the posterior sagittal approach. The orifice of the ACD, measuring 1 to 2 mm in diameter located just behind anus, directed to the lumen of the anal canal by keeping in the midline and ended blindly 5 mm above the dentate line without luminal communication. Histology findings showed a squamous epithelium with smooth muscle bundles in 2 cases and pseudostratified columnar epithelium with focally squamous epithelial lining and adjacent smooth muscle bundles in 3 cases. The postoperative courses were uneventful with satisfactory anal function. One patient has not yet undergone operation and has been well on outpatient follow-up.

Conclusions: ACD is a congenital developmental lesion located in the midline posterior to the anus presenting as a tubular structure without communication with the anal canal, usually discovered in early infancy, and characteristically predominant in girls. The authors recommend that all ACDs, regardless of size and length, should be removed surgically to restore the normal anatomy and to avoid delayed presentation of infection such as perianal abscess or fistula formation.

INDEX WORDS: Duplication of the alimentary tract, anal canal duplication, anal canal.

ANAL CANAL DUPLICATION (ACD) is the most distal digestive duplication located posterior and sagittal to the normal anus and is the least common duplication along the entire alimentary tract. The term *anal canal,* was proposed by Symington[1] in 1888 as a part extending from the level of the pelvic floor to the anal opening and corresponds to the "surgical" anal canal, whereas the term *anatomic anal canal* has been used for the area between the line of anal valves and sinuses (dentate line, DL) and the anal verge alone.[2]

A review of the English-language literature found 15 cases of ACD reported in the pediatric age group. We now present an additional 6 cases of ACD focusing on the clinical characteristics, preoperative imaging, and pathologic findings. We also discuss the embryology of the ACD.

MATERIALS AND METHODS

Between 1999 and 2001, a total of 6 infants who presented with a postanal opening at the 6 o'clock position since birth were seen at the Pediatric Surgery Keimyung University Dongsan Medical Center. The charts were reviewed retrospectively focusing on the clinical characteristics, preoperative diagnosis, and pathologic findings.

Among 6 patients, 5 patients underwent operation, and one has undergone follow-up for 2 years without undergoing operation.

RESULTS

In a short period, ACD was diagnosed in a series of 6 patients. As summarized in Table 1, all patients were girls. A small opening measuring 1 to 2 mm in diameter located just posterior to the anus (Fig 1) in the midline had been noted since birth but ignored. There was no history of erythematous swelling or fluctuation, suggesting perianal abscess, and there was no discharge from the opening. Patients ranged in age from 3 to 9 months at the time of diagnosis (mean, 4.5 months). The anal canal duplication was delineated clearly by contrast study of the tract (Fig 2). All were tubular structures. Ultrasonography or pelvic computed tomography (CT) was used to evaluate other associated anomalies. All patients had an anus in the normal position, a normal sacrum, normal

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Table 1. Cases of ACD

	Case No.					
	1	2	3	4	5	6
Sex	F	F	F	F	F	F
Age at diagnosis (mo)	4	4	4	3	3	9
Age at operation (mo)	5	5	8	6	3	None
Discovery	Inspection	Inspection	Inspection	Inspection	Inspection	Inspection
Site (o'clock)	6	6	6	6	6	6
Type	Tubular	Tubular	Tubular	Tubular	Tubular	Tubular
Length (mm)	12	12	12	10	10	
Digestive communication	None	None	None	None	None	None
Associated anomaly	None	None	None	None	None	None
Diagnosis	Fistulography, US	Fistulography, US	CT	CT, fistulography	None	CT, fistulography
Pathology	Pseudo col squamous	Pseudo col squamous	Squamous	Squamous	Pseudo. Col squamous	None

Abbreviations: US, ultrasound; Pseudo col, pseudostratified columnar epithelium.

anal sphincter tone, and normal anal function. None of the patients had any other associated anomalies. In all cases except one, operation was performed between 3 and 8 months of age (mean, 5.4 months). Excision of the ACD was accomplished through the posterior sagittal approach without difficulty. The course of the ACD remained in the midline and ended blindly 5 mm above the dentate line without communication with the anal canal (Fig 3). The excised specimen was 10 to 12 mm in length. Histology results showed a squamous epithelium with smooth muscle bundles in 2 cases and pseudostratified columnar epithelium with a focally squamous epithelial lining and adjacent smooth muscle bundles in 3 cases (Fig 4). Anal glands were noted in all. The postoperative courses were uneventful with satisfactory anal function.

One patient, first seen at 9 months of age has not yet undergone excision and has been well on outpatient follow-up for 2 years.

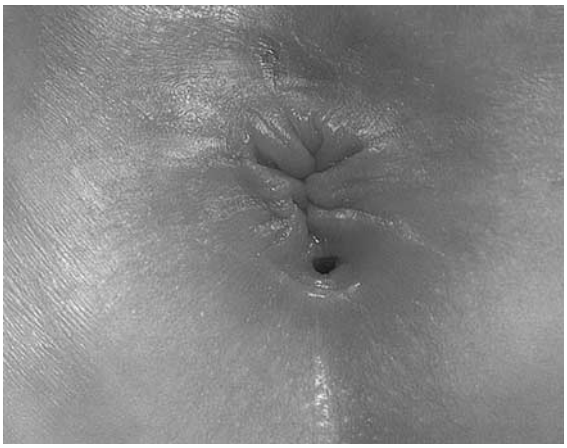

Fig 1. Small opening (measuring 1 mm in diameter) in the posterior midline to anus in case 2.

DISCUSSION

The anal canal is a complex anatomic structure, and the anatomy and embryology are still under debate.[3] The term *anal canal* proposed by Symington[1] in 1888 corresponds to the "surgical" anal canal.[2] The average length of the anal canal is 4.2 cm in a living person and 3 cm in

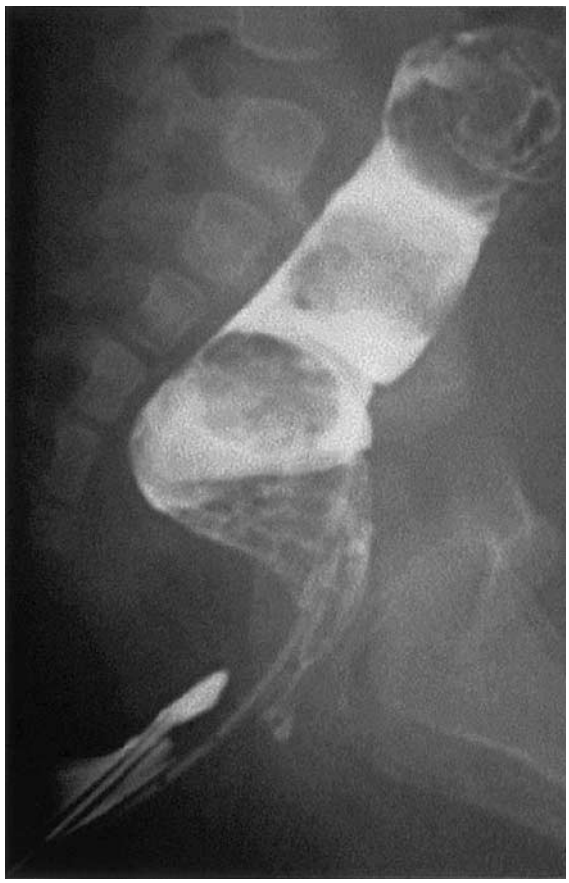

Fig 2. Contrast examination shows tubular structure without communication with the lumen of the normal anal canal in case 2.

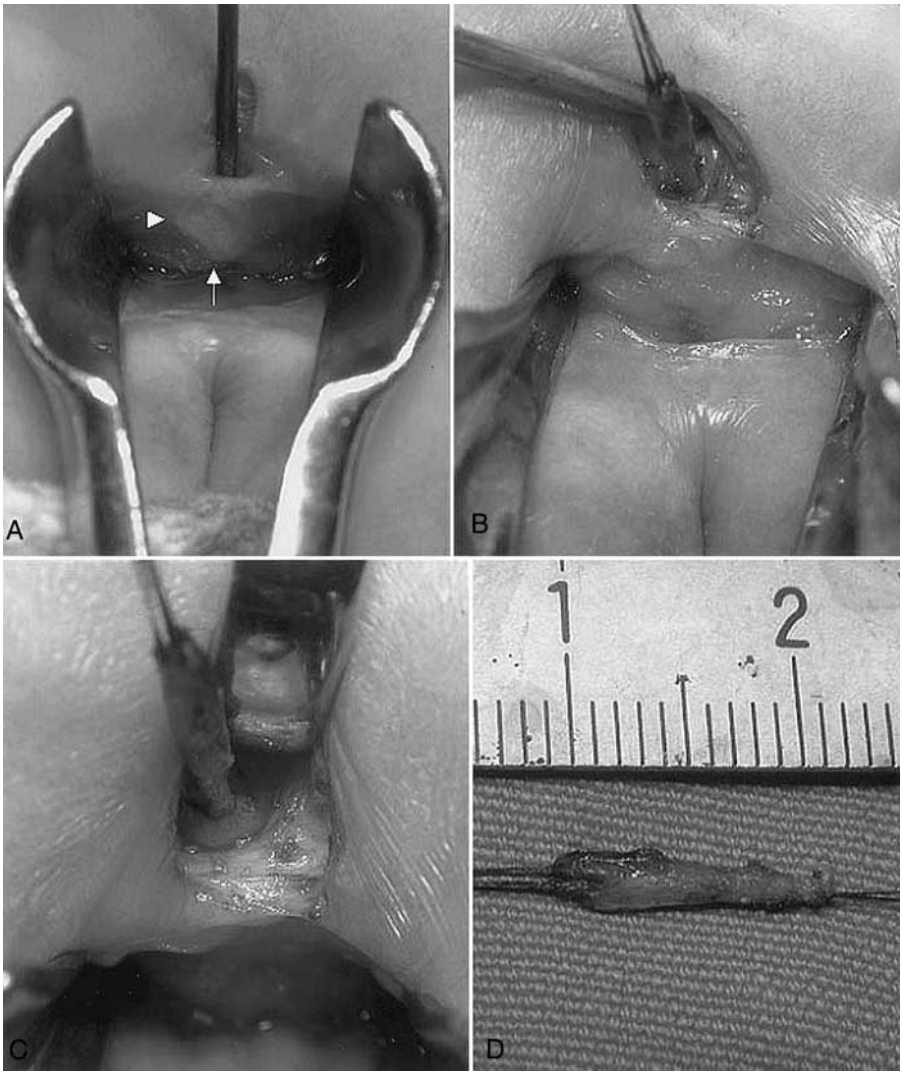

Fig 3. Intraoperative pictures with a probe in the ACD and an anal speculum in normal anus shown blind end of the ACD (white arrow) reached 5 mm above the dentate line (white arrowhead)(A), dissected tract through an incision (B,C), and excised specimen (D) in case 2.

formaline-fixed specimens.[4,5] The anal canal can be divided into 3 zones according to the epithelial lining: colorectal, anal transitional, and squamous.[6] The upper colorectal zone is covered with uninterrupted colorectal mucosa. The anal transitional zone, the zona columnaris, starts at the dentate line (DL), extends 3 to 20 mm cranially, and is covered with epithelial variants (columnar, cuboidal, polygonal or flattened, metaplastic squamous epithelium). The lower squamous zone normally extends downward from the DL and gradually merges into the perianal skin covered with keratinized squamous epithelium and skin appendages.

Anal canal duplication (ACD) is a rare congenital anomaly in which patients present with an external opening just behind the normal anus. The tract runs along the posterior aspect of the anal canal without communication with the anorectum. To date, 15 cases were found to be compatible with the diagnosis of ACD among the 16 cases reported in the English-language literature.[7-11] Four of 5 cases reported as postanal sinus by Ponson and Festen[10] were included, and one caused by an infected dermoid cyst was excluded.

The diagnosis was made at various ages ranging from neonatal period to 12 years. Two-thirds of the patients were diagnosed in the first year of life. Fifteen of 16 patients were girls. ACD is a predominantly tubular structure, and its length is varies from 10 to 30 mm. All girls had the noncommunicating type of ACD including ours, but only one boy, reported by Jacquier et al,[9] had a communicating tubular structure. ACD occurring in boys must be differentiated from the congenital perianal fistula having a fistulous tract to the anal lumen.

ACDs usually are asymptomatic, but early diagnosis can be made by simple perineal inspection, which reveals a small opening located in the midline just behind the normal anus.

The most useful diagnostic modality for identifying the duplication is a contrast study of the tract. The ACD

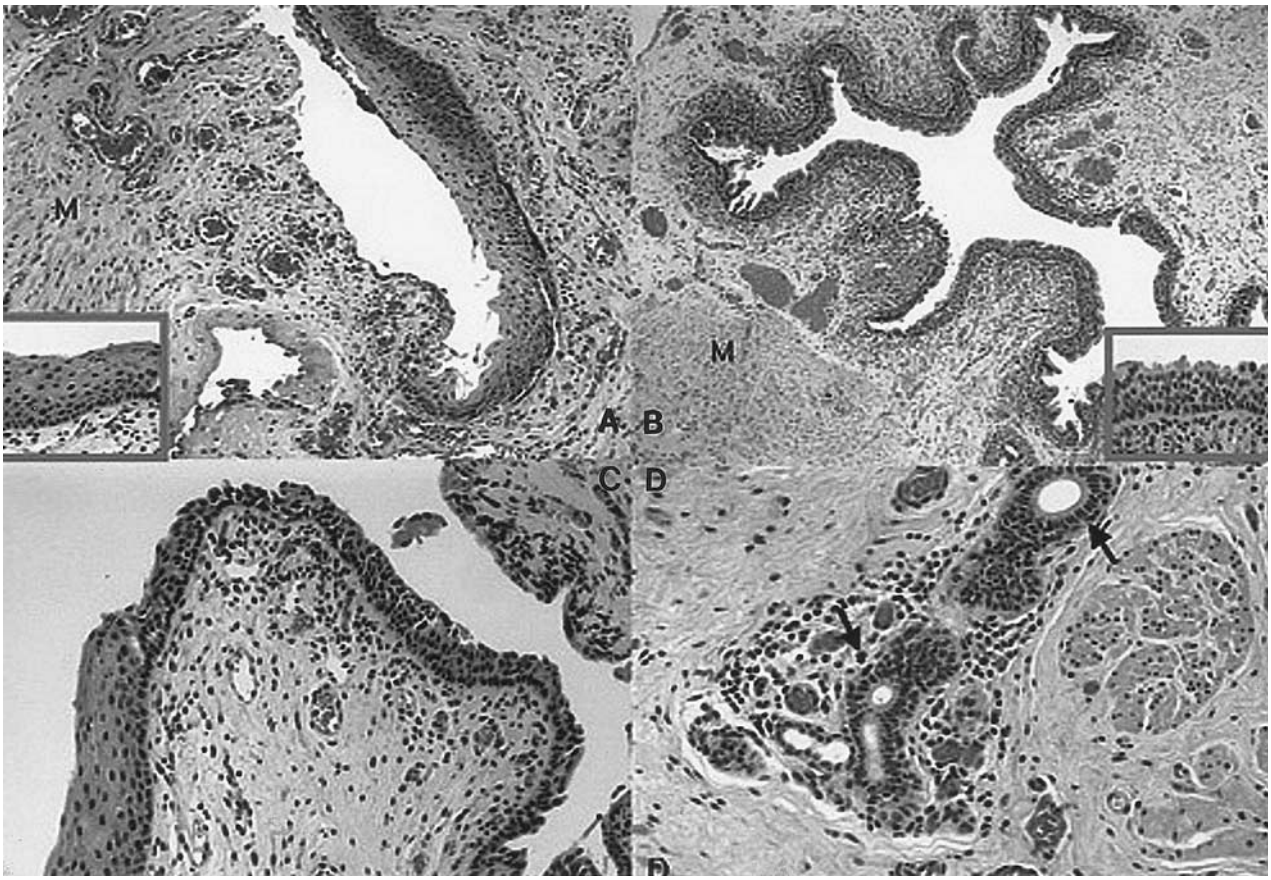

Fig 4. Photomicrographs of the histology of ACDs. Case 4 (A), squamous epithelial lining (inset) with partially denuded and adjacent smooth muscle bundles (M). (H&E, original magnification × 100.) Case 2 (B), pseudostratified columnar epithelial lining (inset) and adjacent smooth muscle bundles (M). (H&E, original magnification × 100.) There is a transition from columnar to squamous epithelium (C) and submucosal anal glands (arrows) are also present (D). (H&E, original magnification × 200.)

is delineated clearly, and more information can be obtained by performing a barium enema of the normal rectum simultaneously. In some cases, ultrasonography, pelvic computed tomography (CT) or pelvic magnetic resonance imaging (MRI) may be necessary to evaluate the presence of associated anomalies, in paticular, sacral dysgenesis, presacral tumor, or hindgut anomaly. Jacquier et al[9] reserve MRI for special cases of a presacral mass. Regarding associated anomalies, dermoid cysts, presacral teratomas, ureteral duplication, lumbosacral meningocele, and spina bifida have been reported.[9-11]

The embryologic basis for ACD with the anus in normal position has not been well established and remains unclear. Hamada et al[8] thought the pathogenesis of ACD might be explained by duplication of the dorsal cloaca in the early developmental stage of the 30-day-old embryo as described by van der Putte.[12] This theory explaines why the ACD is located in the postanal site and is associated with a high rate of concomitant anomalies such as sacral dysgenesis or Currarino's triad. However, more recently, Nievelstein et al[13] described the embryonal development of the anorectum, as occurring through the process of cloacal membrane rupture at the 16 to 18-mm crown-rump length (C-RL) around day 49 of human development and occlusion of the anal orifice with adhesion and formation of a plug of epithelial cells. Recanalization of the anal canal then occurs by apoptotic cell death at the 30 to 35-mm C-RL and remains narrow at least up to the 40-mm C-RL, and eventually an open anal orifice is evident after 40-mm C-RL. A premodial levator ani muscle is recognized around the rectum at the 13 to 17-mm C-RL (around day 44 of human development), but the promyoblast of the external anal sphincter muscle is identified after 40-mm C-RL as a ring of myoblasts at the level of the anal canal. Investigators believe that embryogenesis of ACD with a normal anal position is best explained as a late embryonic defect described by Nievelstein et al.[13] ACD may occur in the fetus that has a greater length of dorsal cloacal membrane than normal. Recanalization of the excess dorsal cloacal membrane after normal development of the external anal sphincter will cause a duplication. This ex-

plains why anal canal duplication develops in the midline posterior to the anus and does not involve the anal sphincter muscle, does not extend to the levator muscle, and does not communicate with the lumen of the anal canal. The course of the ACD in our 5 cases ran along the posterior midline of the anal canal and ended blindly 5 mm above the dentate line without communication with the anal canal.

Hamada et al[8] proposed and defined the term *ACD,* which should be restricted to a single duplication of the anal canal, not including cases of other duplications of the hindgut with or without genitourmary involvement, but including some cases with sacral dysgenesis or congenital anorectal malformations. However, investigators emphasize again that the diagnosis of ACD should be applied to cases of an anal opening in the normal position and a single duplication in the midline posterior to the anus without communication with the anal canal and differentiated from hindgut duplication or congenital fistula in ano.

Histology of the ACD may show a variety of different epithelia according to the length of the duplication. The most common histology was squamous or transitional epithelium with smooth-muscle components and anal ducts around the cavity.[7-11] In none of our patients was rectal mucosa found.

The treatment of ACD is complete excision through the posterior sagittal approach. If the ACD is not excised, it may become infected with age by accumulation of secretions in the lumen that may lead to multiple recurrences and an increase in the size of the orifice by repeated incision and drainage.

The prognosis after complete excision of ACD is good with patients having normal anal function.

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