Pediatric Surgery International (2019) 35:971–978
https://doi.org/10.1007/s00383-019-04509-x

ORIGINAL ARTICLE

Anal duplication: is surgery indicated? A report of three cases and review of the literature

A. C. Trecartin[1] · A. Peña[1] · M. Lovell[2] · J. Bruny[1] · C. Mueller[3] · M. Urquidi[4] · Andrea Bischoff[1]

Accepted: 20 June 2019 / Published online: 29 June 2019

Abstract

Aim of the study Anal canal duplications (ACDs) are extremely rare with only approximately 90 cases described in the literature. We report on three additional patients.

Methods Cases were analyzed to evaluate presenting symptoms, physical exam and MRI findings. A comprehensive literature review was performed to compare our patients to previously described cases. IRB approval was obtained for this study (19-0394).

Main results The first female patient presented with an asymptomatic ACD at 2 years old. The second patient was a 13-year-old female with perianal drainage that was initially mistaken for a fistula-in-ano and ultimately found to have an ACD associated with a dermoid cyst. Both posterior midline duplications shared a common wall with the rectum, but did not communicate with it. The ACDs and dermoid cyst were successfully excised through a posterior sagittal approach with no postoperative complications. Histology demonstrated the presence of both squamous epithelium and transitional anal epithelium in each case. The third patient was 8 months old and had a tethered cord, hemisacrum, presacral mass, and anal duplication that was initially undiagnosed. These results corroborate patterns identified in other reports of ACDs with over 90% being female and in the posterior midline. The majority are asymptomatic, but may present with symptoms of local or even systemic infection.

Conclusion An opening in the midline posterior to the anus should raise clinical suspicion for anal canal duplication. An associated presacral mass must be ruled out. Complete excision through a posterior sagittal approach is recommended upon diagnosis to avoid symptomatic presentations. The key part of the operation is the separation of the ACD from the posterior rectal wall.

Keywords Anal duplication · Presacral mass · Hemisacrum

Background

Anal duplication is a rare condition with only approximately 90 cases described in the English literature. It may be misdiagnosed as a fistula-in-ano, or not identified until symptoms develop [1–3]. Anal duplication is associated with midline anomalies including presacral masses [2, 4]. Anal duplications are so rare that the literature contains only small series of patients. The practitioner has little data with which to guide management decisions. An updated comprehensive literature review was performed to assimilate the currently available case reports and series. We present three cases referred to us and discuss a successful management approach.

✉ Andrea Bischoff
andrea.bischoff@childrenscolorado.org

1 International Center for Colorectal and Urogenital Care, Department of Pediatric Surgery, Children's Hospital Colorado, 13123 East 16th Avenue, Box 323, Anschutz Medical Campus, Aurora, CO 80045, USA

2 Department of Pathology, Children's Hospital Colorado, Aurora, CO, USA

3 Department of Pediatric Surgery, Lucile Packard Children's Hospital, Palo Alto, CA, USA

4 Department of Pediatric Surgery, Clinica Olivos, Cochabamba, Bolivia

Case 1

A 2-year-old female was referred for an anal posterior orifice identified on physical exam (Fig. 1, Patient 1). She had been born full term and had no other medical problems. She was eating normally and had one to two soft formed stools daily. Her sacrum was normal. An MRI of the pelvis demonstrated no presacral mass or spinal abnormality.

She was placed in the prone position for the resection of the anal duplication. An abnormal orifice was visible in the midline posterior to a normal anus. Multiple 5–0 silk sutures were placed in the mucocutaneous junction of the duplication (Fig. 2a). A small posterior sagittal incision through the skin and subcutaneous tissue was performed with needle-tip electrocautery (Fig. 2b). The duplication was dissected circumferentially with meticulous care taken anteriorly where it shared a common wall with the rectum (Fig. 2c). The duplication was noted to be a blind pocket measuring 1.5 cm in length. This was excised in its entirety and sent to pathology. She continues to stool normally and had no complications.

Pathology noted that the specimen consisted of a narrow stellate lumen lined completely by stratified squamous epithelium (Fig. 2d) except for a focal area of transitional epithelium in the deep aspect. The lumen had a blind end. There was no inflammation. There were small clusters of mucinous glands surrounded by disorganized bundles of smooth muscle within interlacing fibrovascular tissue as confirmed by the trichrome stain (Fig. 2e). There were no ganglion cells. The histology was consistent with an anal canal duplication in the posterior midline.

Case 2

A 13-year-old female presented with several weeks of sacral pain. She had normal bowel function and reported normal soft stools every other day. She then developed purulent drainage from her perineum and her mother noted an abnormal opening behind the anus. She was started on antibiotics for a presumed fistula-in-ano. Pediatric surgery evaluated the patient and identified an anal duplication (Fig. 1, Patient 2). Her sacral radiograph was normal. An MRI demonstrated a 3.4 × 2.9-cm complex fluid collection in the presacral space and right ischiorectal fossa (Fig. 3a, b).

A posterior sagittal approach was performed, dividing skin, subcutaneous tissue, and the sphincter complex in the posterior midline. The tract was meticulously dissected circumferentially and was found to share a common wall with the rectum. This was dissected free and the posterior rectal wall left completely intact. This epithelialized anal duplication tract was associated with a 3.5 × 3-cm cystic mass. The mass was entirely excised and it was verified that there were no other masses. A drain was placed. The posterior sphincter mechanism was sutured in layers with 4–0 Vicryl placing special emphasis on suturing corresponding structures. The drain was removed on postoperative day number 6. The patient continued to stool normally and had no infection. Follow-up MRI 6 months later demonstrated no residual abnormality.

Histology identified a lumen lined by transitional anal epithelium at the distal end. A single deep mucinous gland was noted in the adjacent fibrovascular soft tissue. One cyst lined by squamous epithelium contained neutrophils, macrophages, and necrotic keratin debris (Fig. 3d). Hair shafts within inflamed soft tissue were identified. At the proximal

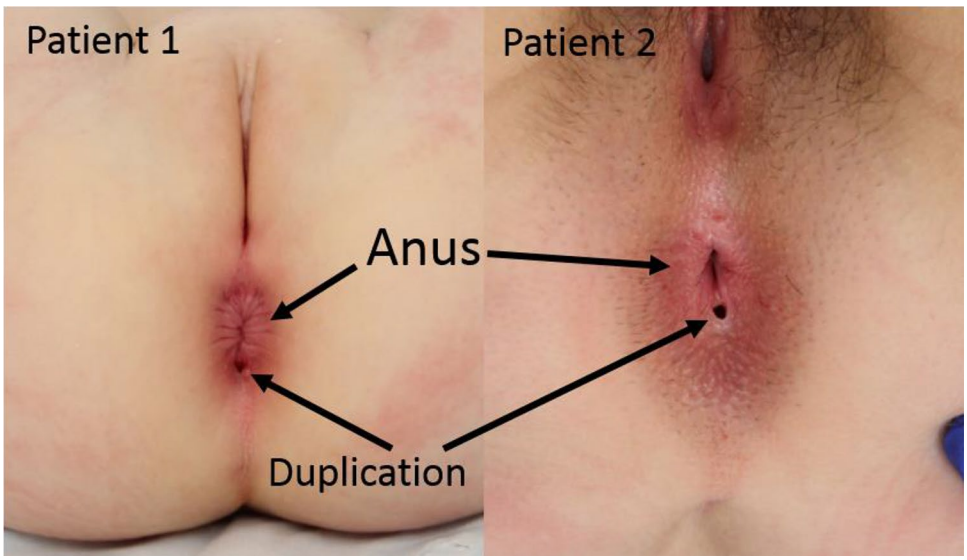

Fig. 1 The anal duplication is clearly identified in the posterior midline for patients 1 and 2

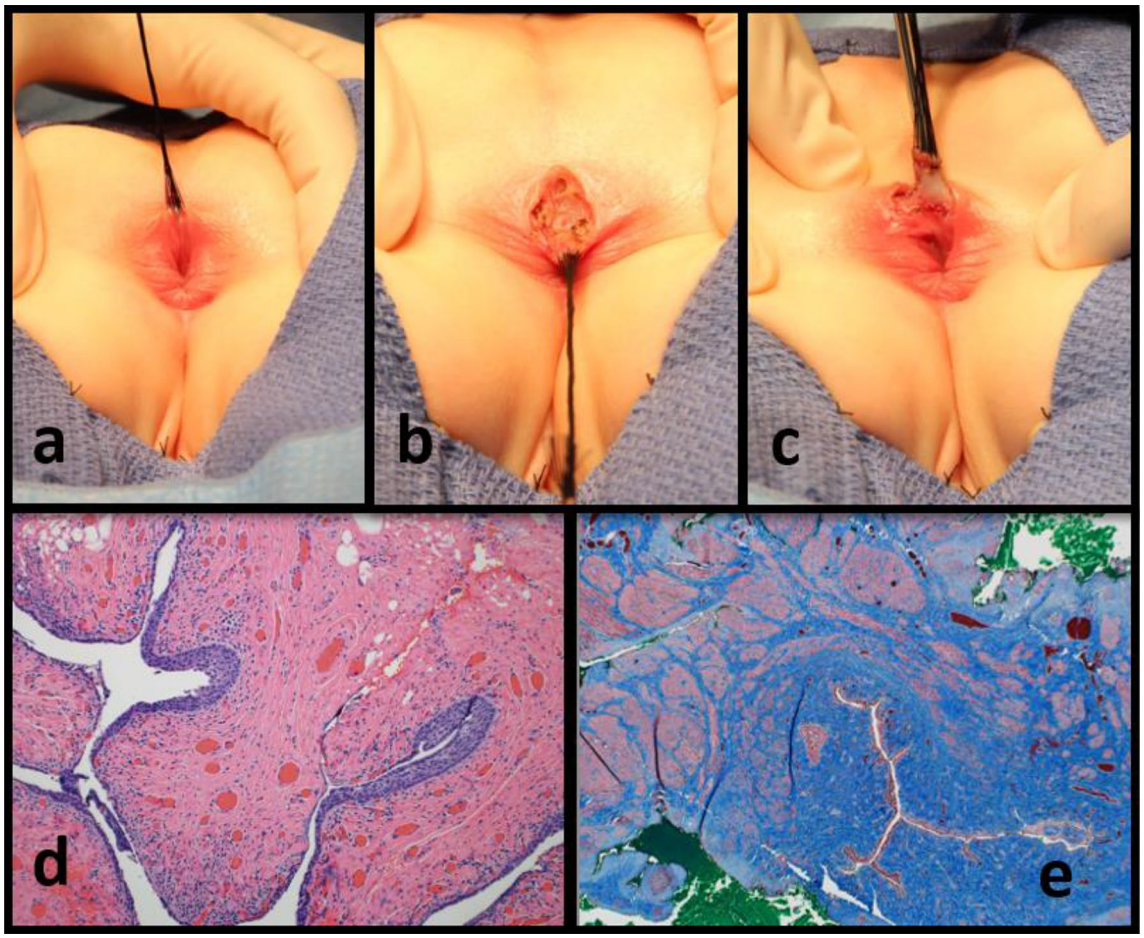

Fig. 2 Patient 1 is in the prone position with multiple silk traction sutures placed at the muco-cutaneous junction circumferentially around the anal duplication (**a**). A small posterior sagittal incision is performed (**b**). Anteriorly, the duplication shares a common wall with the rectum which must be meticulously separated (**c**). The lumen was lined by stratified squamous epithelium (**d**) and trichrome stain highlights fibrovascular tissue surrounding disorganized bundles of smooth muscle (**e**)

end of the specimen, several cystic spaces lined by keratinizing squamous epithelium were seen (Fig. 3e). Slips of smooth muscle were seen surrounding the anal duplication and cyst. Histopathology was consistent with an anal canal duplication and ruptured dermoid cyst.

Case 3

An 8-month-old female in Bolivia was noted to have an anorectal malformation with a perineal fistula (Fig. 4a). Her workup revealed a tethered cord, hemisacrum (Fig. 4b), presacral mass, single vagina, normal right kidney, left pelvocaliectasis, no reflux on voiding cystogram, and an anorectal duplication. A colostomy had been created at birth and the senior author was invited to perform her reconstruction.

A posterior sagittal approach facilitated complete resection of the 5-cm presacral mass (Fig. 4c) and anal duplication. A posterior sagittal anorectoplasty was also performed. There were no postoperative complications.

Literature review

A comprehensive literature review was performed to identify patterns in the cases reported to date. Cases with sufficient available details are included in Table 1. One of the earliest and largest series of anal duplications was of 11 patients reported by Dr. Hardy Hendren in 1990 [5]. The concern for confusing an anal duplication with a fistula-in-ano was discussed at that time and remains relevant as in our second patient. Nine additional cases of anal canal duplications have been reported with fewer available pertinent details [6–8]. An early report of a similar condition was described in 1956 associated with colloid carcinoma; however, the columnar epithelial lining was more consistent with rectum and unlike the stratified squamous epithelium of the anal canal [9]. In 1970, a case series included

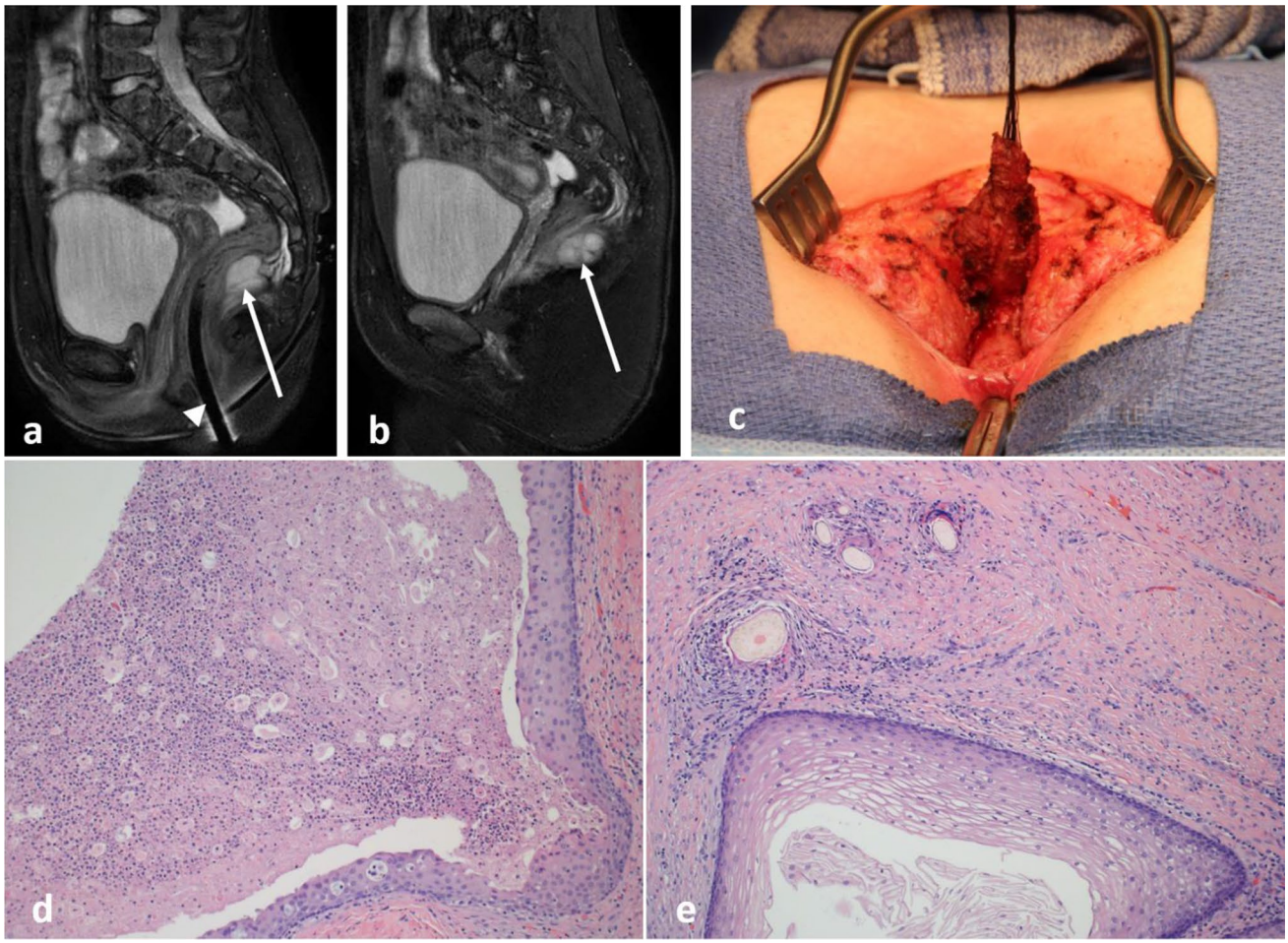

Fig. 3 Patient 2 with a Peña–Patel protocol MRI (**a**, **b**) showing the catheter in the rectum (arrowhead). It identifies the cystic presacral mass (white arrows). Silk traction sutures were placed on the anal duplication together with the mass (**c**). Note the Hegar dilator in the rectum and intact posterior rectal wall (**c**). A ruptured dermoid cyst is shown (**d**) and the keratinizing squamous epithelium lining the anal duplication (**e**)

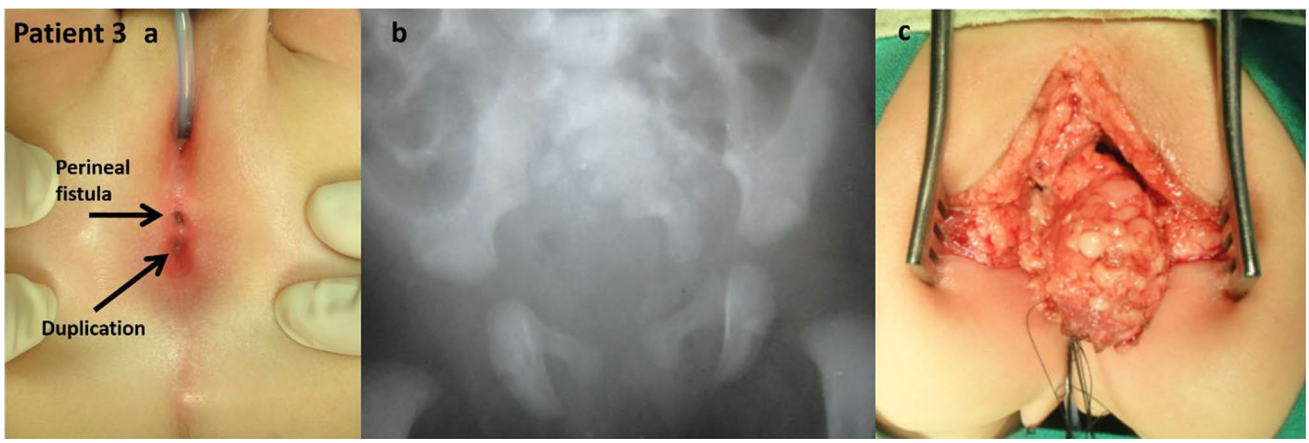

Fig. 4 Patient 3: the anal canal duplication was identified posterior to the anorectal malformation with perineal fistula (**a**). The patient also had a hemisacrum (**b**) indicating a presacral mass shown in **c**

Table 1 Reports of anal duplications with focus on presenting symptoms and additional anomalies.

Report	Number of patients	Age at presentation	Sex	Symptoms	Associated anomalies	Location of opening(s)
Honda et al. (2017) [1]	1	14 years	F	Noticed at 1 y/o, but asymptomatic until developed abscess	None	Posterior midline
Palazon et al. (2017) [8]	8	2 months 2 years 5 years 6 years 10 years 8 years 18 months 10 years	F ($n=6$) M ($n=2$)	Asymptomatic ($n=6$), constipation ($n=1$), painful anal mass ($n=1$)	None	Posterior midline except for one anal triplication patient. Two patients had two openings each in the posterior midline except one was 1.5 cm posterior and to the right
Koga et al. (2010) [2]	10	1 month 4 years 8 months 4 years 24 days 2 months 1 month 10 months 7 months 1 year	F	Constipation ($n=7$), infected ($n=1$), meningitis ($n=1$)	Hypoplastic kidney ($n=1$), anal stenosis ($n=2$), tethered cord together with presacral teratoma ($n=1$)	All posterior midline
Sinnya et al. (2013) [3]	1	15 years	F	Perianal pain, fevers	Infected anal duplication cyst	Posterior midline
Pampal et al. (2013) [11]	2	3 months 2 years	F	Anorectal malformations ($n=2$)	Ureteric duplication with vesicoureteral reflux, spina bifida, sacral and hip abnormalities ($n=1$)	Posterior midline
Van Biervliet et al. (2013) [12]	1	11 years	F	Anal pruritus	None	Posterior midline
Cheng et al. (2013)	1	1 year	F	Asymptomatic	none	Posterior midline
Mirzaei et al. (2015) [13]	4	20, 50, 25, and 24 years	F	Perianal abscess/fistula, perineal mass, discharge, 1 asymptomatic	Perineal cystic masses ($n=2$)	One anterior opening, one lateral opening
Carpentier et al. (2009) [4]	2	3 months 2 months	F M	Sepsis from infected presacral mass, tethered cord ($n=1$), malodorous drainage ($n=1$)	Presacral cystic mass ($n=1$)	Posterior midline (one communicated with anal canal)
Kratz et al. (2008) [14]	1	16 years	F	Lower abdominal pain	Presacral cystic mass	Posterior midline
Tiryaki et al. (2006) [15]	2	7,7 years	F	Asymptomatic, abdominal pain, mucous drainage	Intrasacral meningocele ($n=1$)	Posterior midline
Choi et al. (2003) [16]	6	Mean 4.5 months (range 3–9 months)	F	Asymptomatic	None	Posterior midline
Ochiai et al. (2002) [17]	1	6 years	F	Pruritus	Presacral mass	Posterior midline

Table 1 (continued)

Report	Number of patients	Age at presentation	Sex	Symptoms	Associated anomalies	Location of opening(s)
Lisi et al. (2006) [18]	12	At birth (n = 3) 8 months 27 months 30 months 60 months 2 months 41 months 19 months 3 months 24 months	F (*n* = 11) M (*n* = 1)	Asymptomatic (*n* = 6) Constipation (*n* = 3), recurrent fistula (*n* = 2), mucous discharge (*n* = 1)	Presacral mass (*n* = 3) (2 mature teratoma), IND type B with presacral ependymoma, ARM with recto-vestibular fistula (*n* = 1), lumbar hemangioma, cleft palate with cloacal extrophy variant (*n* = 1)	All posterior midline except 1 not recorded and 1 just to the right of the posterior midline. One cystic mass had no visible opening
Ponson et al. (2001) [19]	5	10 years 1 year 4 years 10 months	F	Asymptomatic (*n* = 2) (1 incidentally had hematochezia), anal pain with discharge (2), discharge alone (1)	Dermoid cyst (*n* = 1), infected dermoid cyst (*n* = 1)	All posterior midline
Jacquier et al. (2001) [20]	6	5 < 1 year old	F (*n* = 5) M (*n* = 1)	Asymptomatic (*n* = 5), perineal pain and diarrhea (*n* = 1)	Presacral teratoma (*n* = 2) (1 also had myelomeningocele)	All posterior. One case communicated with the anal canal
Hamada et al. (1996) [21]	2	7 months, 6 years	F	Asymptomatic	Cleft lip (*n* = 1)	Posterior midline
Arai et al. (1990) [22]	2	7 days, 2 years	F	Anal pain with constipation (*n* = 1), vomiting (*n* = 1)	Not reported	Posterior midline
Lippert et al.(2011) [23]	1	12 years	F	Anal pain, pressure, fevers	Infected presacral mass (*n* = 1)	Posterior midline
Narci et al. (2010)	2	9 months 1 year	F	Asymptomatic	None	Posterior midline
Vietan et al.(2009) [24]	1	3 months	F	Asymptomatic	None	Posterior midline

the word anal duplication; however, on closer evaluation these were a family of three patients with anorectal malformations, sacral defects, and presacral masses without any description of a true anal duplication [10]. A total of 71 patients with available pertinent details are included in Table 1.

Discussion

The clinician must suspect an anal duplication when examining an orifice posterior to a normal anus. The diagnosis requires evaluation for a presacral mass. Surgical excision is recommended to prevent development of symptoms.

Almost all anal duplications open in the posterior midline and occur in females as in our cases [2, 20]. The majority have no additional abnormalities. However, many will have midline anomalies that should not be missed [25]. Concomitant anomalies include myelomeningocele, presacral teratoma or other presacral mass, congenital heart defects, cleft lip/palate, genitourinary anomalies, and tethered cord. Regarding prognosis for bowel control, patient 2 had bowel control pre-operatively and remains with bowel control post-operatively, patient 1 is still on diapers, and due to the presence of presacal mass and hemisacrum, we do not expect patient 3 to achieve bowel control. Bowel control should be expected in patients with isolated anal duplications [16].

Except for rare cases, the anal canal duplication does not communicate with the rectum [8]. Our patients did not demonstrate any fistula between the duplication and the native rectum. A key finding, however, is that the anal duplication is intimately associated with the posterior rectal wall making meticulous surgical dissection critical.

Surgical resection is indicated to prevent future symptoms and potential malignancy. The standard surgical approach is posterior sagittal with excellent outcomes [25]. Some reports describe a mucosectomy without complete excision due to concern for damaging the anal sphincter [15]. We recommend complete excision through a posterior sagittal incision to reduce the risk of injury to the posterior rectal wall, or incomplete excision. A small posterior sagittal incision allows full visualization for complete excision. When performed correctly, the posterior sagittal approach does not risk any injury that would cause incontinence. Care must be taken to avoid injury to the posterior rectal wall as the anal duplication is immediately adjacent to and may share a common wall with the rectum.

Conclusion

The clinician should suspect an anal canal duplication when visualizing an orifice posterior to the anus. An associated presacral mass must be ruled out. Complete excision is recommended to avoid symptomatic presentations. A posterior sagittal approach facilitates safe dissection. The critical part of the operation is meticulous separation of the anal canal duplication from the posterior rectal wall.

References

1. Honda S, Minato M, Miyagi H, Okada H, Taketomi A (2017) Anal canal duplication presenting with abscess formation. Pediatr Int 59(4):500–501
2. Koga H, Okazaki T, Kato Y, Lane GJ, Yamataka A (2010) Anal canal duplication: experience at a single institution and literature review. Pediatr Surg Int 26(10):985–988
3. Sinnya S, Curtis K, Walsh M, Wong D, Kimble R (2013) Late presentation of anal canal duplication in an adolescent female—a rare diagnosis. Int J Colorectal Dis 28(8):1175–1176
4. Carpentier H, Maizlin I, Bliss D (2009) Anal canal duplication: case reviews and summary of the world literature. Pediatr Surg Int 25(10):911–916
5. La Quaglia MP, Feins N, Eraklis A, Hendren WH (1990) Rectal duplications. J Pediatr Surg 25(9):980–984
6. Tagart RE (1977) Congenital anal duplication: a cause of para-anal sinus. Br J Surg 64(7):525–528
7. Hata YSK (1978) Duplication of the rectum and/or anal canal. Jpn J Pediatr Surg 14:105–109
8. Palazon P, Julia V, Saura L et al (2017) Anal canal duplication and triplication: a rare entity with different presentations. Pediatr Surg Int 33(5):609–617
9. Dukes CE, Galvin C (1956) Colloid carcinoma arising within fistulae in the anorectal region. Ann R Coll Surg Engl 18(4):246–261
10. Aaronson I (1970) Anterior sacral meningocele, anal canal duplication cyst and covered anus occurring in one family. J Pediatr Surg 5(5):559–563
11. Pampal A, Ozbayoglu A, Kaya C et al (2013) Rectal duplications accompanying rectovestibular fistula: report of two cases. Pediatr Int 55(4):e86–89
12. Van Biervliet S, Maris E, Vande Velde S et al (2013) Anal canal duplication in an 11-year-old-child. Case Rep Gastrointest Med 2013:503691
13. Mirzaei R, Mahjubi B, Alvandipoor M, Karami MY (2015) Late presentation of anal canal duplication in adults: a series of four rare cases. Ann Coloproctol 31(1):34–36
14. Kratz JR, Deshpande V, Ryan DP, Goldstein AM (2008) Anal canal duplication associated with presacral cyst. J Pediatr Surg 43(9):1749–1752
15. Tiryaki T, Senel E, Atayurt H (2006) Anal canal duplication in children: a new technique. Pediatr Surg Int 22(6):560–561
16. Choi SO, Park WH (2003) Anal canal duplication in infants. J Pediatr Surg 38(5):758–762
17. Ochiai K, Umeda T, Murahashi O, Sugitoh T (2002) Anal-canal duplication in a 6-year-old child. Pediatr Surg Int 18(2–3):195–197
18. Lisi G, Illiceto MT, Rossi C, Broto JM, Jil-Vernet JM, Lelli Chiesa P (2006) Anal canal duplication: a retrospective analysis of 12 cases from two European pediatric surgical departments. Pediatr Surg Int 22(12):967–973
19. Ponson AE, Festen C (2001) Postanal sinus: single or different etiologies? Pediatr Surg Int 17(1):45–47
20. Jacquier C, Dobremez E, Piolat C, Dyon JF, Nugues F (2001) Anal canal duplication in infants and children: a series of 6 cases. Eur J Pediatr Surg 11(3):186–191
21. Hamada Y, Sato M, Hioki K (1996) Anal canal duplication in childhood. Pediatr Surg Int 11(8):577–579

22. Arai T, Miyano T, Tanno M, Kohno S, Hamasaki Y (1990) Tubular anal duplication: experiences with two cases. Z Kinderchir 45(5):311–313
23. Lippert SJ, Hartin CW Jr, Ozgediz DE (2012) Communicating anal canal duplication cyst in an adolescent girl. Colorectal Dis 14(5):e270–271
24. Vietan D, Patel RV, Huddart SN (2009) Duplicated proctodeum. Pediatr Surg Int 25(6):535–536
25. Cheng LS, Courtier J, MacKenzie TC (2013) Anal duplication in a one-year-old girl. J Pediatr Surg Case Rep 1(10):373–374