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Pediatric Surgical Image

A call to ARMs: Accurate identification of the anatomy of the rectourethral fistula in anorectal malformations

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ABSTRACT

Recto-urethral fistulae are the most common finding in males born with an anorectal malformation (ARM). A high pressure distal colostogram is an important tool in visualizing the fistula, although the precise level at which the fistula communicates with the urethra may be difficult to interpret and is not reported in a uniform manner. This anatomy affects the surgical plan; determining the likelihood that the rectum is reachable via a posterior sagittal incision or better approached through the abdomen via laparoscopy or laparotomy helps counsel families and stratifies diagnoses for outcomes work. Herein we present a figure used at our Center to assist with correlating findings on the distal colostogram with the anatomic level of the recto-urethral fistula in males with anorectal malformations.

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1. Background

Fistulae to the urinary tract are the most common findings in males born with an anorectal malformation. The anatomy distributes across a spectrum of disorders with wide ranging complexity which affects the surgical management, rate of associated anomalies, and long-term functional outcomes [1]. As such, the term "rectourethral fistula" is imprecise and therefore insufficient in conveying important information necessary for surgical planning, counseling of families, and expected long-term functional outcome.

High pressure distal colostogram is the most accurate diagnostic study in determining the presence and level of a rectourethral fistula in males, but these studies may be difficult to interpret with precision [2,3]. Herein we present a figure which we use for educational and clinical purposes to correlate findings on the distal colostogram with identification of the precise location of the rectal fistula (recto-bulbar, recto-prostatic, or recto-bladder neck) in males born with an anorectal malformation.

2. Presentation

The right arm of the sculpture shown in Fig. 1 is used to depict the course of the male urethra from the bladder as seen on distal colostogram. We correlate levels of the sculpture's arm to clinically relevant urethral levels consistent with the modern classification of anorectal malformations [4] [Fig. 1A]. The elbow or below represents the bulbar urethra [Fig. 1B], the humerus represents the prostatic urethra [Fig. 1C], and axilla represents the bladder neck [Fig. 1D]. The level at which the fistula communicates with the urethra defines the specific ARM subtype as either a recto-bulbar, recto-prostatic fistula, or recto-bladder neck fistula.

In 86 of 86 male patients repaired at our Center between 2014 and 2018, use of this figure correctly diagnosed the rectourethral anatomy. Of these, there were 38 bulbar, 26 prostatic, and 22 bladder neck fistulae. The fistula level was confirmed intraoperatively in all cases. This information obtained in the preoperative period facilitated appropriate operative planning, either via PSARP or laparoscopy in all cases.

3. Discussion

The early management of males born with an anorectal malformation and an absent perineal anal opening focuses on identification and management of associated anomalies included in the

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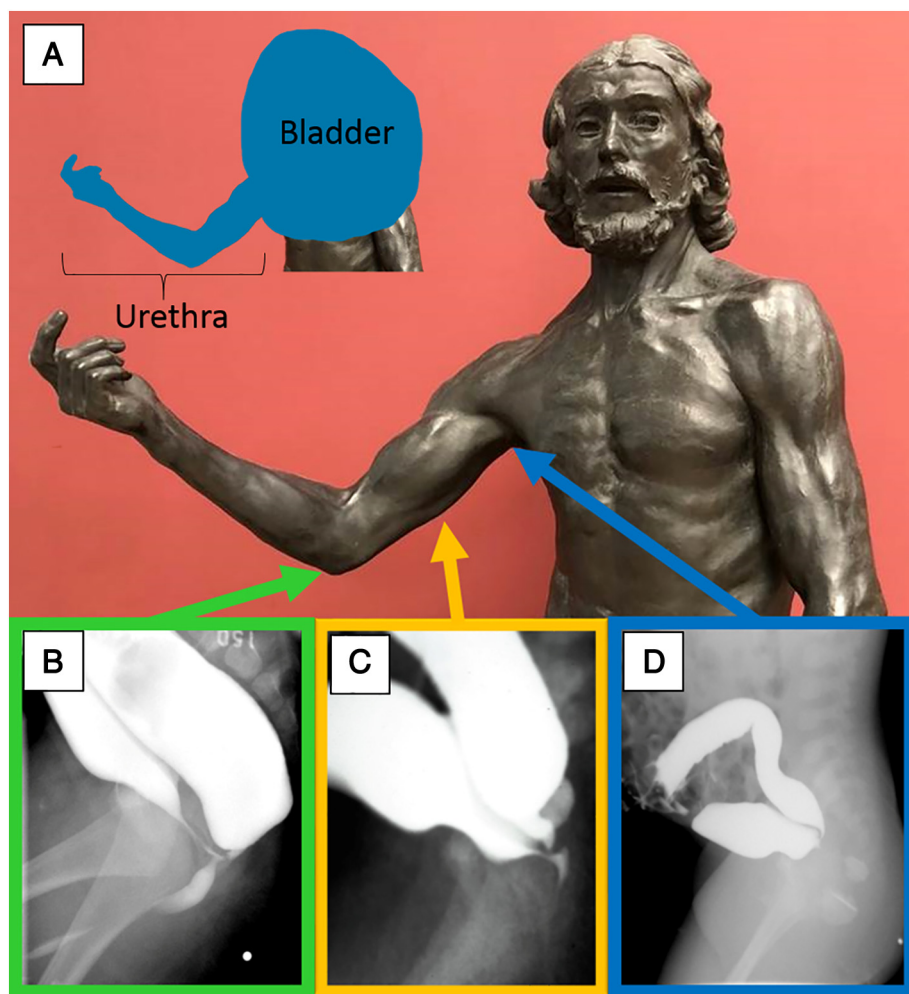


Fig. 1. Auguste Rodin's Saint John the Baptist sculpture (1880) in the Musée d'Orsay, Paris, France [9]. The right arm is used to represent the course of the male urethra (A). The relative positions of the male urethra are represented by the various levels of the arm of the statue. The elbow represents the bulbar urethra (B), the humerus represents the prostatic urethra (C), and the axilla represents the bladder neck (D).

VACTERL association followed by fecal diversion in the first 48 h of life. Thereafter, high-pressure distal colostogram performed through the mucous fistula is performed to study the distal rectal anatomy [2]. If clinical suspicion for a rectal fistula exists despite a negative distal colostogram, then a voiding cystourethrogram can be an important adjunct to further delineate the anatomy. The early categorization of ARMs as either "high," "intermediate," or "low" malformations was replaced by the modern classification system that utilizes the precise anatomic location of the distal rectal fistula [4,5].

Clinicians often describe these patients as having a "recto-urethral fistula," but we believe a more precise assessment is needed. A correct anatomic definition facilitates surgical planning by informing the surgeon whether the rectum can be reached from a posterior sagittal incision only, or whether a dissection via the abdomen will be needed as well [Fig. 2]. For example, a bladder neck fistula frequently requires the addition of laparotomy or laparoscopy, while a bulbar fistula can usually be repaired by a posterior sagittal incision alone. This information is also useful in determining the expected length of the case, what operative equipment will be needed, what prep will be required, strategies for effective postoperative pain, and predicted length of hospital stay.

Additionally, the height of the rectum is associated with fecal continence later in life [1,6,7], and therefore a precise understanding of the level of the fistula is needed in order to properly counsel families about their child's expected continence potential. More complex malformations are associated with lower rates of fecal continence, and thus this knowledge can be used to inform parents about the likelihood that their child will require bowel management options as either a bridge to continence or as a long-term solution to keeping their child clean and in normal underpants [1].

Lastly, describing the level of rectal fistula is crucial to allow for benchmarking of outcomes across institutions by comparing similar anatomic cases to each other and thus permits the accurate study of long-term functional outcomes at a multi-center level [8].

4. Conclusion

A precise description of the level of rectal fistula in male patients born with an anorectal malformation assists in surgical planning, counseling of families, and benchmarking to allow for ongoing study of outcomes. The diagram presented herein allows for the simple and reliable identification of the fistula level in males with rectourethral fistulas.

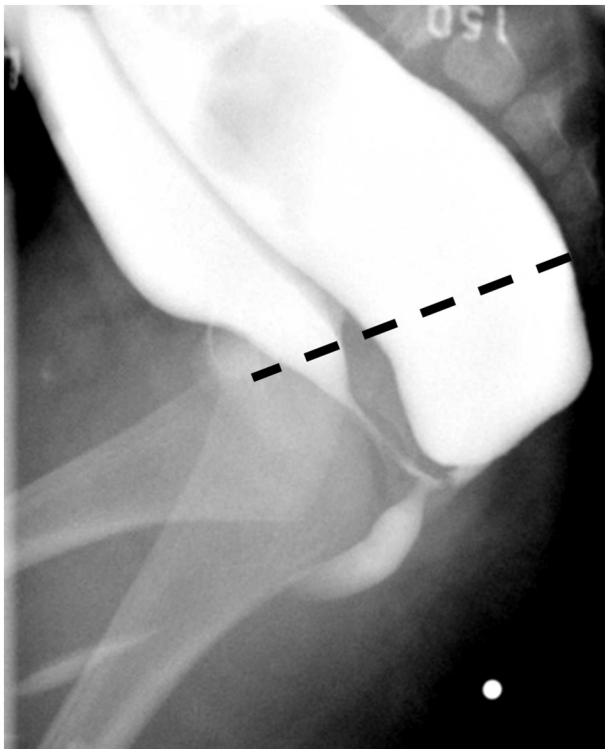


Fig. 2. Distal colostogram of a male patient born with an anorectal malformation. The pubococcygeal line (dotted line) may be useful in predicting the need for laparoscopy or laparotomy at the time of posterior sagittal anorectoplasty. If the rectum is the first structure encountered from a posterior sagittal approach, then the case can more likely be performed using only a posterior sagittal approach. If the urinary tract is the first structure that would be encountered via a posterior sagittal incision, then it is more likely that laparoscopy or laparotomy likely be needed.

References

- [1] Levitt MA, Pena A. Anorectal malformations. *Orphanet J Rare Dis* 2007;2:33.
- [2] Kraus SJ, Levitt MA, Pena A. Augmented-pressure distal colostogram: the most important diagnostic tool for planning definitive surgical repair of anorectal malformations in boys. *Pediatr Radiol* 2018;48(2):258–69.
- [3] Gross GW, Wolfson PJ, Pena A. Augmented-pressure colostogram in imperforate anus with fistula. *Pediatr Radiol* 1991;21(8):560–2.
- [4] Holschneider A, Hutson J, Pena A, et al. Preliminary report on the international conference for the development of standards for the treatment of anorectal malformations. *J Pediatr Surg* 2005;40(10):1521–6.
- [5] Stephens FD, Durham Smith E. Classification, identification, and assessment of surgical treatment of anorectal anomalies. *Pediatr Surg Int* 1986;1(4):200–5.
- [6] Tannuri AC, Ferreira MA, Mathias AL, et al. Long-term evaluation of fecal continence and quality of life in patients operated for anorectal malformations. *Rev Assoc Med Bras* 2016;62(6):544–52.
- [7] Minneci PC, Kabre RS, Mak GZ, et al. Can fecal continence be predicted in patients born with anorectal malformations? *J Pediatr Surg* 2019. <https://doi.org/10.1016/j.jpedsurg.2019.02.035>.
- [8] Reeder RW, Wood RJ, Avansino JR, et al. The pediatric colorectal and pelvic learning consortium (PCPLC): rationale, infrastructure, and initial steps. *Tech Coloproctol* 2018;22(5):395–9.
- [9] Rodin A. St. John the Baptist preaching. Paris, France: Musée d'Orsay; 1880.