Congenital Anterior Urethrocutaneous Fistula: A Systematic Review

Yang Lin, Changkai Deng, Qiang Peng

Department of Pediatric Surgery, Chengdu Women's and Children's Central Hospital of Chongqing Medical University, Chengdu, China

Abstract

Congenital anterior urethrocutaneous fistula (CAUF) is a rare anomaly characterized by fistulization of penile urethra to skin. It's usually seen as an isolated deformity or may accompany genitourinary or anorectal malformations. We aim to define the common properties of patients mentioned in literatures by systematic review. A comprehensive search of PubMed, Embase, Web of Science, and Cochrane Library was performed including cross-referencing independently by two assessors. Selections were restricted to human studies in English. Based on the systematic review, 63 patients in 34 articles were included in the study. Most common fistula site was subcoronal in 29 (46.0%) patients. Chordee was in 8 (14.5%) and associated genitourinary anomaly was detected in 19 (30.2%) of patients. Fistula recurrence ratio was 6/59 (11.3%) using different surgical techniques and 3/6 was closed spontaneously. CAUF is frequently located in subcoronal level and usually an intact urethra distal to it. Success rates are high with the principles of hypospadias surgery.

Keywords: Children, congenital, fistula, surgery, urethra

Introduction

Congenital anterior urethrocutaneous fistula (CAUF) is an uncommon anomaly of the penile urethra. It's usually seen as an isolated deformity or may accompany genitourinary or other malformations.[1-3] The cause is unclear but probably reflects a focal defect in the urethral plate that prevents fusion of the urethral folds.[4,5] Over the past few decades, more and more cases were reported. However, the information gathered about this condition mostly comes from case reports and a few original articles with limited number of patients. To overcome the limitation of individual studies, we carried out this systematic review to provide a more precise and comprehensive properties of CAUF.

Methods

Four databases (PubMed, Web of Science, Embase, and Cochrane Library) were electronically searched to retrieve studies on CAUF by September 10, 2017. Searching terms were ("urethral" AND "fistula" AND "congenital") OR ("urethrocutaneous fistula" AND "congenital"). In additional, we evaluated all associated publications to identify the most eligible literature. Their reference lists were hand-searched to obtain other relevant publications. This systematic review was based on the preferred reporting items for systematic reviews and meta-analyses' guidelines.

Inclusion criteria and exclusion criteria

Titles and abstracts of all relevant papers were reviewed first. Then, full texts were reviewed as a second screening. The studies were considered eligible if they met all of the following criteria: (i) the study explored CAUF; (ii) when multiple publications reported on the same or overlapping data, the most recent article or the article based on the largest study population was selected; (iii) the publication language was English. Studies met any of the following exclusion criteria were excluded: (i) researches based on animals or cells rather than general population; (ii) reviews, editorials, meeting abstracts, and commentaries; and (iii) articles with no target data or no relevant outcomes.

Access this article online

Quick Response Code:

Website: www.afrjpaedsurg.org

DOI: 10.4103/ajps.AJPS_97_17

Address for correspondence: Dr. Changkai Deng, Department of Pediatric Surgery, Chengdu Women's and Children's Central Hospital of Chongqing Medical University, No. 1617, Riyue Avenue, Qinyang District, Chengdu, China.
E-mail: 295969893@qq.com

How to cite this article: Lin Y, Deng C, Peng Q. Congenital anterior urethrocutaneous fistula: A systematic review. Afr J Paediatr Surg 2018;15:63-8.

Two reviewers (Changkai Deng and Yang Lin) reviewed all eligible publications independently according to the aforementioned inclusion and exclusion criteria; then, we extracted the relevant data in accordance with the preformed data extraction form. Disagreements were solved by discussion and a third party (Qiang Peng) was involved when necessary. The basic information was extracted from each article: first author, year of publication, country where study was conducted, sample size of case, history of circumcision, location of fistula, associated urinary system anomalies, associated other anomalies, type of surgical repair, number of recurrence, number of catheter days, and auxiliary examination.

Results

We initially identified 996 potentially eligible studies. Most of them were excluded after the screening of titles and abstracts. The main excluded reason was duplication and irrelevant to CAUF. After assessing the full-text of 44 potentially relevant articles, we identified 34 eligible articles. The main reasons for exclusion were as follows: five studies were no relevant outcome, three studies were commentaries, and two studies reported on the same data [Figure 1].

Thirty-four studies with 63 cases were included in the analysis. The basic characteristics of included studies are presented in Table 1. All studies were published between 1962 and 2017. The included studies were conducted in India, USA, UK, Turkey, France, Italy, and so on. Subcoronal fistula was detected in 29 patients, midpenile in 24, proximal penile to subcoronal level in 4, and penoscrotal in 1. Eleven cases were circumcised. Fistula recurrence ratio was 6/59 and 3/6 was closed spontaneously. Eight cases with chordee, 19 cases with associated genitourinary anomalies, and 11 cases with associated other anomalies. Complicated cases were 19 and isolated cases were 44.

Discussion

CAUF is defined as a localized defect in penile urethra of congenital origin. It's usually seen as an isolated disorder or may accompany genitourinary or other penile anomalies. Limited information exists about this topic since it is a rare anomaly and the clinical characteristics are not properly defined. The etiology of CAUF is still obscure, and several

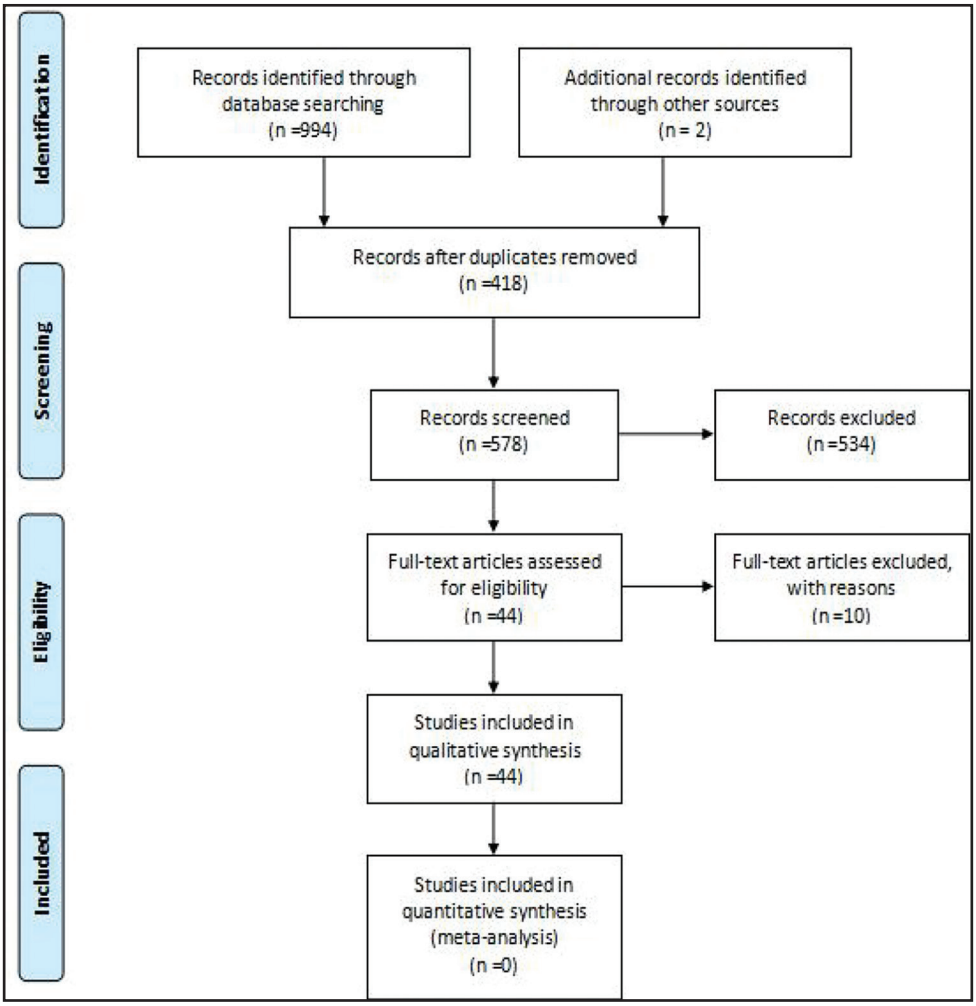

Figure 1: Flow diagram depicting the search strategy

Table 1: Characteristic of the included studies

First authour	Year	Isolated/ complicated	Country or region	Case number	History of circumcision	Location of fistula	Chordee
Gupta AK	2017	Complicated	India	1	No	Penoscrotal	No
Raj P	2017	Complicated	India	1	No	From proximal penile to subcoronal level	No
User IR	2016	Isolated	Turkey	3	No	Midpenile (2)/ subcoronal (1)	No
Cheng SY	2016	Complicated	China-Taiwan	1	No	Penoscrotal	No
Kale SM	2015	Isolated	India	1	No	Scrotum	No
Biswas S	2014	Isolated (8)/ complicated (1)	India	9	No	Subcoronal (7), Midpenile (2)	1
Alhazmi HH	2014	Isolated	Saudi Arabia	2	1	Subcoronal	No
Spinelli C	2013	Isolated	Italy	1	No	Midpenile	No
Bhatnagar A	2012	Isolated	India	2	No	Subcoronal (1), Midpenile (1)	No
Kumara TL	2011	Isolated	Sri Lanka	1	No	From proximal penile to subcoronal level	No
Jindal T	2011	Isolated	India	1	1	Subcoronal	No
Shukla RM	2011	Isolated	India	3	No	Midpenile (2), Penoscrotal (1)	No
Chen Q	2011	Isolated	China	1	No	Midpenile	No
Galinier P	2009	Complicated	France	1	No	From proximal penile to subcoronal level	No
Rashid KA	2008	Isolated	India	1	1	Subcoronal	No
Arena F	2008	Isolated	Italy	1	Null	Midpenile	No
Ceylan K	2006	Complicated	Turkey	1	No	Midpenile	1
Akman RY	2005	Isolated	Turkey	1	No	Midpenile	No
Merrot T	2003	Complicated	France	1	No	Midpenile	No
Islam MK	2001	Isolated	Bangladesh	1	No	Midpenile	No
Nakane A	2000	Complicated	Japan	1	No	Midpenile	1
Harjai MM	2000	Isolated	India	1	No	Midpenile	No
Sharma AK	2000	Isolated	India	1	No	Midpenile	No
Caldamone AA	1999	Isolated (8)/ complicated (6)	USA	14	5	Subcoronal (10), Midpenile (2), Penoscrotal (2)	2
Maarafie A	1997	Isolated	UK	1	No	Midpenile	No
Barwell J	1997	Complicated	UK	1	No	Midpenile	1
Karnak I	1995	Isolated	Turkey	1	No	Subcoronal	No
Ritchey ML	1994	Complicated	USA	1	1	Midpenile	No
Tennenbaum SY	1994	Isolated	USA	2	2	Subcoronal	No
Olbourne NA	1976	Isolated (1)/ complicated (1)	UK	2	No	Subcoronal	No
Goldstein M	1975	Complicated	USA	1	No	Midpenile	1
Shiraki IW	1973	Complicated	USA	1	1	From proximal penile to subcoronal level	1
van der Meulen JC	1971	Isolated	Netherlands	1	Null	Midpenile	No
Gupta MS	1962	Isolated	UK	1	No	Subcoronal	No

Location of urethral meatus	Corpus spongiosum distal to fistula	Associated urinary system anomalies	Associated other anomalies	Type of surgical repair	Recurrence	The number of catheter days
No	Intact	YUD	ARM/urethrorectal fistula	Vaginalis flap	No	Null

Contd...

Table 1: Contd...

First authour		Year	Isolated/ complicated	Country or region	Case number	History of circumcision	Location of fistula	Chordee
No	Intact	Bilat. undescended testes	Type V pouch colon (ARM)/prune belly syndrome	Repair the CAUF at later date			Null	Null
Tip of glans (1)	Intact (2)/ deficient (1)	No	No	3-layer closure			1 (resolved after regular dilatation fo meatus)	7
No	Intact	Megalourethra	No	Referred to a specialist for an optimal surgical correction			Null	Null
No	Intact	No	No	Dartos flap			No	14
No	Intact	No	ARM (1)	Bayer's flap (7), Tunica vaginalis flap (2)			No	Null
No	Intact	No	No	Dartos flap			1 (was closed spontaneously with 3 months)	7
No	Intact	No	No	Preputial onlay flap			No	7
No	Intact	No	Left inguinal hernia (1)	Local tissue flaps in two layers (1), Snodgrass (1)			No	Null
No	Intact	No	ARM	3-layer closure/dartos flap			No	5
No	Intact	No	No	2-layer closure			No	Null
No	Intact	No	ARM (1)	3-layer closure/dartos flap/local tissue flaps			No	Null
No	Intact	No	No	3-layer closure			No	Null
No	Intact	Stenotic bulbar urethra/ bifid scrotum/ penoscrotal transposition	ARM	Conventional surgical technique			No	Null
No	Intact	No	No	Tiersch-Duplay			No	Null
No	Intact	No	No	3-layer closure			No	6
No	Intact	No	No	Transverse preputial island flap			No	Null
No	Intact	No	No	Transverse preputial island flap			No	9
No	Intact	Solitary left kidney/ duplicated urethral/ reflux in a blind-ending rignt ureter	No	Unspecified			No	Null
No	Intact	No	No	Primary repair with local tissue flap			No	9
No	Intact	No	No	Transverse preputial onlay island flap			No	Null
No	Intact	No	No	Preputial onlay flap/3-layer closure			No	5
No	Intact	No	No	2-layer closure			No	10
No	Intact (6)/ deficient (8)	VUR (1)/ undescended testes (3)	Cong heart disease (1)/ ARM (3)	Tiersch-Duplay (6), Snodgrass (2), Mathieu (2), Preputial onlay flap (2), Island pedicle interpositiontube (2)			2	Null
No	Intact	No	No	Preputial vascularized onlay flap with hypospadias repair			Null	Null
No	Deficient	No	Bilateral inguinal hernia	Urethral reconstruction with split preputial flap			No	Null
No	Intact	No	No	Proximal-based skin flap			1	Null
No	Intact	Hypoplastic left kidney/ bilat. undescended testes/ hypospadias	PDA/dextrocardia/ARM	3-layer closure			No	3
No	Intact	No	No	Tiersch-Duplay/dartos flap			No	Null

Contd...

Table 1: Contd...

First authour		Year	Isolated/ complicated	Country or region	Case number	History of circumcision	Location of fistula	Chordee
No	Intact	Diverticulum	No	Denis Browne procedure/3-layer closure			1 (was closed spontaneously with 3 months)	Null
No	Intact	Hypoplastic right kidney	ARM/rectoprostatic fistula	Proximal-based skin flap+transverse preputial island flap			No	7
No	Intact	Megalourethra	No	Primary repair with preputial flap			No	No
No	Intact	Null	Null	Null			Null	Null
No	Intact	No	No	Denis Browne procedure			No	Null

ARM: Anorectal malformations, PDA: Patent ductus arteriosus, YUD: Y-type urethral duplication, CAUF: Congenital anterior urethrocutaneous fistula

pathogenetic theories have been used to explain its causes.[5-7] Olbourne suggested that fistulae located in the penile shaft probably reflect a focal or temporary defect in urethral plate function and this would result in a complete defect or a partial deficiency of urethral fold fusion.[7] Goldstein theorized that a transient deficiency in testicular evocator substance could produce congenital urethral fistula with chordee.[6] Karnak regarded congenital urethrocutaneous fistulas (excluding those associated with anorectal malformations) as one set of anomalies.[5]

Based on systematic review, 63 patients in 34 articles were included since 1962. Most of the articles report a few number of cases except 2 articles cover 14 and 9 cases, respectively.[8,9] Subcoronal fistula was detected in 29 patients, midpenile in 24, proximal penile to subcoronal level in 4, and penoscrotal in 1. Subcoronal and midpenile locations seem to be typical for CAUF. Presentation of patients after circumcision raises the question whether this fistula might be acquired caused by the procedure or congenital and it becomes obvious after circumcision. Eleven of 63 patients reported in the literature were circumcised, and in some cases, the fistula is located proximal to prepuce making it unlikely to be iatrogenic.[4,9-13]

Associated penile and urethral anomalies may be encountered in these patients as complicated cases. Eight patients with chordee and 19 with associated genitourinary anomalies were reported up to date with CAUF cases.[1,6-9,13-20] Meanwhile, associated other anomalies may be detected in patients with CAUF. Based on our systematic review, anomalies which may be noticed by physical examination such as undescended testes (n = 5), inguinal hernia (n = 2), penoscrotal transposition (n = 1), bifid scrotum (n = 1), duplicated urethra (n = 1), megalourethra (n = 2), anorectal malformation (n = 11), and congenital heart disease (n = 2) were detected in cases mentioned in the literature. This indicated that it's better to advise a through physical examination in all patients and these additional anomalies may necessitate different surgical techniques in fistula repair and make the procedure more complicated.[2]

The management of these anterior urethrocutaneous fistulae depends on the size and location. Small fistulas which are <0.5 cm can be easily closed primarily after refreshing the edges and covered by skin.[21] Fistulas of size >0.5 cm but <1 cm can be closed with turnover flap.[3,6] Larger fistulas which are >1 cm can be closed by tubularized incised plate urethroplasty using the Thiersch-Duplay technique.[1,9,12,20] Fistulae larger than 2 cm are associated with significant defect in urethra and skin cover which can be dealt with bilamellar preputial island flap.[22] Based on our systematic review, success rates are high with all the principles of hypospadias surgery. Fistula recurrence ratio was 6/59 (11.3%) using different surgical techniques and 3/6 (50.0%) was closed spontaneously.[2,5,7,9,10]

Conclusion

This systematic review suggested that CAUF is chiefly located in subcoronal level and usually an intact urethra distal to it and treatment of this entity is personalized according to site of fistula, associated anomalies, and condition of the distal urethra. Success rates are high with all the principles of hypospadias surgery.

Financial support and sponsorship

Nil.

Conflicts of interest

Yang Lin and Changkai Deng contributed equally to this work.

References

1. Gupta AK, Kumar M, Singh K, Sokhal AK. Rare association of congenital penile urethrocutaneous fistula with Y-type urethral duplication. BMJ Case Rep 2017;2017. pii: bcr2016217331.
2. User IR, Karakus SC, Akçaer V, Özokutan BH, Ceylan H. Congenital anterior urethrocutaneous fistula: 3 new cases and review of literature. Arch Esp Urol 2016;69:238-43.
3. Akman RY, Cam K, Akyuz O, Erol A. Isolated congenital urethrocutaneous fistula. Int J Urol 2005;12:417-8.
4. Ritchey ML, Sinha A, Argueso L. Congenital fistula of the penile urethra. J Urol 1994;151:1061-2.
5. Karnak I, Tanyel FC, Hiçsönmez A. Congenital urethrocutaneous fistula: A case report and literature review, with a nomenclature proposal. J Pediatr Surg 1995;30:1504-5.
6. Goldstein M. Congenital urethral fistula with chordee. J Urol 1975;113:138-40.
7. Olbourne NA. Congenital urethral fistula. Case reports. Plast Reconstr Surg 1976;57:237-9.
8. Biswas S, Ghosh D, Das S. Congenital urethrocutaneous fistula-our experience with nine cases. Indian J Surg 2014;76:156-8.

9. Caldamone AA, Chen SC, Elder JS, Ritchey ML, Diamond DA, Koyle MA, *et al.* Congenital anterior urethrocutaneous fistula. J Urol 1999;162:1430-2.
10. Alhazmi HH. Congenital anterior urethrocutaneous fistula: Two case reports and review of literature. Urol Ann 2014;6:239-41.
11. Jindal T, Kamal MR, Mandal SN, Karmakar D. Isolated congenital urethrocutaneous fistula of the anterior urethra. Korean J Urol 2011;52:368-70.
12. Rashid KA, Kureel SN, Tandon RK. Congenital anterior penile isolated urethrocutaneous fistula: A case report. Afr J Paediatr Surg 2008;5:52-3.
13. Shiraki IW. Congenital megalourethra with urethrocutaneous fistula following circumcision: A case report. J Urol 1973;109:723-6.
14. Raj P, Birua H. Type V pouch colon, prune belly syndrome, and congenital anterior urethrocutaneous fistula. J Neonatal Surg 2017;6:38.
15. Cheng SY, Chen SJ, Lai HS. Congenital anterior urethrocutaneous fistula at the penoscrotal junction with proximal penile megalourethra: A case report. J Radiol Case Rep 2016;10:33-7.
16. Galinier P, Mouttalib S, Carfagna L, Vaysse P, Moscovici J. Congenital anterior urethrocutaneous fistula associated with a stenosis of the bulbar urethra in the context of high anorectal malformation without fistula. J Plast Reconstr Aesthet Surg 2009;62:e11-3.
17. Ceylan K, Köseoğlu B, Tan O, Atik B. Urethrocutaneous fistula: A case report. Int Urol Nephrol 2006;38:163-5.
18. Merrot T, Pankevych T, Chaumoitre K, Alessandrini P. Congenital anterior urethrocutaneous fistula associated with urethral duplication. Pediatr Surg Int 2003;19:744-5.
19. Nakane A, Hayashi Y, Kojima Y, Mizuno K, Okada A, Sasaki S, *et al.* Congenital urethrocutaneous fistula. Int J Urol 2000;7:343-4.
20. Barwell J, Harris D. Case report: Congenital urethrocutaneous fistula. J Anat 1997;190(Pt 1):155-6.
21. Shukla RM, Mukhopadhyay B, Mandal KC, Barman SS. Congenital penile urethrocutaneous fistula: A rare anomaly and review of literature. Urol Ann 2011;3:161-3.
22. Islam MK. Congenital penile urethrocutaneous fistula. Indian J Pediatr 2001;68:785-6.