

**A congenital anterior urethrocutaneous fistula in a  
boy whose mother was exposed to ionizing  
radiations: a case report and literature review**

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## Abstract

Anterior congenital urethrocutaneous fistula is a rare anomaly that may present in an isolated fashion or in association with other anomalies of the genital urinary tract or anorectal malformations.

A case of congenital anterior urethrocutaneous fistula non associated with other congenital anomalies in a 3 year old male whose mother was been exposed to Chernobyl's nuclear fallout is described. The patient was successfully operated with no recurrence. We report a review of the literature about etiology and surgical strategy including the role of ionizing radiations.

The congenital anterior urethrocutaneous fistula represents a rare malformation. The etiopathogenesis is unknown.

## Introduction

Primary anterior urethrocutaneous fistula is a very rare malformation with only 28 cases reported in English – language literature from 1962 to 2012 [1-20]. This anomaly is frequently associated with anorectal malformation or other anomalies of the genital - urinary tract, such as hypospadias, chordee or cryptorchidism. It has been demonstrated that ionizing radiation can be the cause of congenital urogenital malformation.

We report an additional case of isolated congenital anterior urethrocutaneous fistula discussing its etiology as well as reviewing the literature.

## Case presentation

A 3 year old, Italian - Ukrainian, not circumcised male came to our attention for the presence of a fistula located on his ventral side of the penis since birth. He had no history of trauma or surgical intervention and his family history did not included hereditary diseases for genital - urinary tract malformations, but the mother was exposed to the nuclear fallout that followed the explosion of the Chernobyl nuclear power plant when she was 1 year old. The systemic and local physical examination of the patient was, except from the fistula, normal; the preputial skin was intact, both testicles were in place and chordee was absent as well as other anomalies. On the ventral side of the midshaft of the penis an opening measuring 5 x 3 mm was present (Fig. 1). It was 2 cm distal to penoscrotal junction and easily passed by a 8 F urethral catheter. The urine was passing mostly through the fistula but also from the external urethral meatus. Investigations such as urine - analysis, abdominal ultrasound and cystourethrography were normal. So we decided to operate the boy. The fistula was closed by one - stage transverse preputial onlay island flap urethroplasty over the 8 F tube that was kept in place for 7 days (Fig. 2). After a 12 months follow - up, the child is healthy without recurrence.

## Discussion

Anterior urethrocutaneous fistula without anorectal malformation or other anomalies of the genital - urinary tract, such as hypospadias, chordee or cryptorchidism, is an extremely rare anomaly with just 28 cases previously reported in literature (Tab. 1) [1-20].

The etiology of congenital urethrocutaneous fistula is not clear yet as well as the exact development of the male urethra. In male fetuses, sexual differentiation and urethral development approximately begin at the<sup>th</sup> week of development when, under the stimulation of testosterone,

the urethral folds begin to close along the midline of ventral surface of the penis. Throughout a similar process, the proximal portion of the granular urethra form shortly thereafter and it is thus derived from the urethral plate, endodermal origin. According to the classical theory, the distal portion of the granular urethra is formed by lamellar ingrowths of the surface epithelium, ectodermal origin, which grows toward the distal extent of urethral plate, becoming stratified squamous epithelium around the 15<sup>th</sup> week. However this theory for the development of the distal granular urethra has been challenged by the "endodermal differentiation theory". It has been suggested that the entire penile urethra might differentiate from the fusion of epithelial - mesenchymal interactions [21].

The variability of the patients reported leads to several pathogenetic theories of the congenital urethrocutaneous fistulas development. Campbell et al. [22] proposed that congenital urethrocutaneous fistulas represent embryonal urethral blowouts behind a distal congenital obstruction. Olbourne et al. [2] suggested that focal defect in the urethral plate result in arrested distal migration of the urethral plate or a localized deficiency of a portion of the plate, which prevents fusion of the urethral folds. Goldstein et al. [23] theorized that there is a transient deficiency or inhibition of the local effect of testosterone leading to the failed closure of the urethral groove. Cook and Stephens [24] suggested an alternative mechanism that should be considered, namely, pressure atrophy from the heel of the baby's foot, leading to pressure necrosis. Karnak et al. [5] regarded congenital urethrocutaneous fistulas (excluding those associated with anorectal malformations) as one set of anomalies. Finally, in circumcised patient, possible urethral injuries can be the cause of an acquired urethrocutaneous fistula [24].

Due to the frequent association of congenital urethrocutaneous fistulas with hypospadias and other genitor - urinary tract anomalies, some genetic pathogenetic theories have been proposed. Genetics studies proved that mutations of ATF 3 (Activating Transcription Factor 3), an estrogens responsive gene expressed during genital development, SHH (sonic Hedge Hog), FGF 8 and 10 (Fibroblast Growth Factors), Ephrin - B2 and receptors EphB2, EphB3, often associated with genitourinary anomalies are also implied in the genesis of isolated urethrocutaneous fistulas [26-29]. Moreover, it has been reported that environmental hazards like ionizing radiation can cause genitor-urinary tract anomalies, reinforcing the hypothesis concerning the cause-effect relationship between the presence of the CAUF in our patient and the mother's exposure to ionizing radiation. [30,31]

According to literature, several surgical techniques for the repair of isolated anterior urethrocutaneous fistula exist including pedicle flap, prepuce bound skin flap, modified Denis Brown urethroplasty or direct closure. The method of repair must be decided on the bases of the location and the size of the fistula and on the presence of other anomalies such as hypospadias or chordee [24]. One of the most important aspect to evaluate is the urethra beyond the fistula. If it is congenitally defective, the simple closure is usually unsuccessful and the fistula is likely to recur. If adequate lateral skin is present, repair can be carried out as one wound, performing a second - stage Johanson urethroplasty [25]. Otherwise, an onlay flap can be also used. In our case the fistula was an isolated abnormality and the urethra beyond the fistula was intact so a primary closure could have been possible if the dimension of the defect was inferior. This is the reason why we preferred the transverse preputial island flap to repair the lesion; another reason was also because the flap seems to maintain a stable, well vascularised thick adequately durable tissue to cover the lesion.

The congenital anterior urethrocutaneous fistula represents, as our work and the literature review demonstrate, a rare malformation. The etiopathogenesis is unknown and the case we described is particularly interesting because the mother was exposed, during the pediatric age, to the nuclear fallout that followed the explosion of the Chernobyl nuclear power plant; this opens an interesting and extensive discussion on the possible correlation between the isolated urethrocutaneous fistula and the exposure to ionizing radiations.

**Consent**

Written informed consent was obtained from the patient's legal guardian for publication of this manuscript and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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	Author	Treatment	N° case	N° recurrence
1962	Gupta <sup>1</sup>	Denis-Browe	1	-
1996	Olbourne <sup>2</sup>	Denis-Browe	2	2
1994	Ritchey <sup>3</sup>	Three layers closure	1	-
1994	Tenenbaum <sup>4</sup>	Thiersch Duplay	2	-
1995	Karnak <sup>5</sup>	Proximal based skin flap	1	1
1997	Barwell <sup>6</sup>	Preputial Flap	1	-
1997	Maarafie <sup>7</sup>	Preputial Flap	1	-
1999	Caldamone <sup>8</sup>	Thiersch Duplay	6	-
2000	Nakane <sup>11</sup>	Preputial Flap	1	-
2000	Sharma <sup>9</sup>	Two layers closure	1	-
2000	Harjai <sup>10</sup>	Preputial Flap	1	-
2003	Betalli <sup>12</sup>	Three layers closure	1	1
2004	Agarwal <sup>13</sup>	Local sksn turndown flap	1	-
2005	Akman <sup>14</sup>	Preputial Flap	1	-
2006	Ceylan <sup>15</sup>	Preputial Flap	1	-
2008	Rashid <sup>16</sup>	Thiersch Duplay	1	-
2008	Arena <sup>17</sup>	Three layers closure	1	-
2009	Spinelli	Preputial Flap	1	-
2009	Galinier <sup>18</sup>	Bladder mucosal graft	1	-
2011	Jindal <sup>19</sup>	Two layers closure	1	-
2011	Qiang <sup>20</sup>	Local skin turndown flap	1	-
			<b>Total</b>	<b>Total</b>
			28	4

**Table 1:**  
Literature  
review



**Figure 1:** Congenital anterior urethrocutaneous fistula before surgery.



**Figure 2:** The ventral surface of the penis after immediately surgical correction by one - stage transverse preputial onlay island flap urethroplasty.