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Median raphe deviation, association with hypospadias and clinical significance: A case report and literature review

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ABSTRACT

Here, we present a case of hypospadias and median raphe deviation with literature review. A 4-year-old patient presented with subcoronal hypospadias and treated by using tubularized incised plate urethroplasty technique. Additionally, a longitudinal dorsal dartos flap was harvested and transposed to the ventral side by the buttonhole maneuver. The ventral view of the patient penis showed a 15 degree deviation to the right side of the center.

Key Words: Hypospadias; median raphe deviation; Snodgrass technique; dorsal dartos flap.

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Introduction

Hypospadias is defined as more proximal placement of the urethral meatus penile than the glans type in the ventral region [1,2]. Embryologically, it can be described as a development defect with urethral spongiosum and ventral prepisum anomalies and the correction of normal penile curvature. It is the most common genital anomaly that have been seen in 1/300 of boys and characterized by ventral opening of the urethral meatus, abnormal penile curvature and abnormal penile skin distribution [1,2]. According to Duckett's classification, Urethral opening of the glans can be divided into subcoronal, distal, midpenil, proximal, penoscrotal, scrotal and perineal. The most common forms are glenoid and subcoronal [1-3]. Metal advancement urethroplasty (MAGPI), glans approximation procedure (GAP), Mathieu's procedure (meatal based flap) and Snodgrass method are commonly used in cases of distal hypospadias (glandular, subcoronal) [4]. Fistula formation is the most common complication [5,6], and various procedures have been described to prevent it [7,8].

The raphe of the penis is defined as a narrow, dark streak or ridge continuous posteriorly with the raphe of the scrotum and extending forward along the midline on the ventral surface of the penis [9]. The median raphe abnormalities such as hyperpigmentation, deviation, division, cyst, canal and sinus can be isolated or sometimes associated with severe congenital genitourinary malformations. They

can detected in any period of life. It is important to note that the presence of raphe abnormalities leads to functional, infectious and aesthetic complications [10]. Deviation of the penoscrotal raphe from the normal midline position is associated with hypospadias and angle of glanular torsion [9,11]. Here, we present a case of hypospadias and median raphe deviation with literature review.

Case report

The 4-year-old patient's subcoronal hypospadias was seen at examination as applied for circumcision purposes by his parents. The patient had no recurrent urogenital and endocrinological illnesses before the operation.

At the local examination of the patient, the mea was located at the coronal level [Fig. 1A]. The ventral view of the patient penis showed a 15 degree deviation to the right side of the center [Fig. 1B]. The patient was operated under general anesthesia.

After the urethra was placed in the catheter, an incision was made from subcoronal and ventral prepisum to buck's fascia. The skin and the dartos fascia were separately degloved with dorsal pedicle. After the cord excision, a urethral meatus was created with new urethral tubularization (Snodgrass technique). The dorsal dartos fascia was separated from both the skin and the mucosa and the proximal pedicle dorsal dartos fascia was prepared [Fig. 1C]. A hole was made 1.5 cm in the middle of the fascial flap [Fig. 1D]. The glans penis was passed through this hole and the fascia was tumbled [Fig. 1E] and placed as a second layer over the ventral-formed new urethra [Fig. 1F]. The dorsal skin was incised from the middle into two proximal pedicle flaps and the flaps were shifted from their sides to the ventricle and the left flap was positioned proximally and right flap distally as a skin cover and the preputial excesses were excised with circumcision. The incisions were sutured.

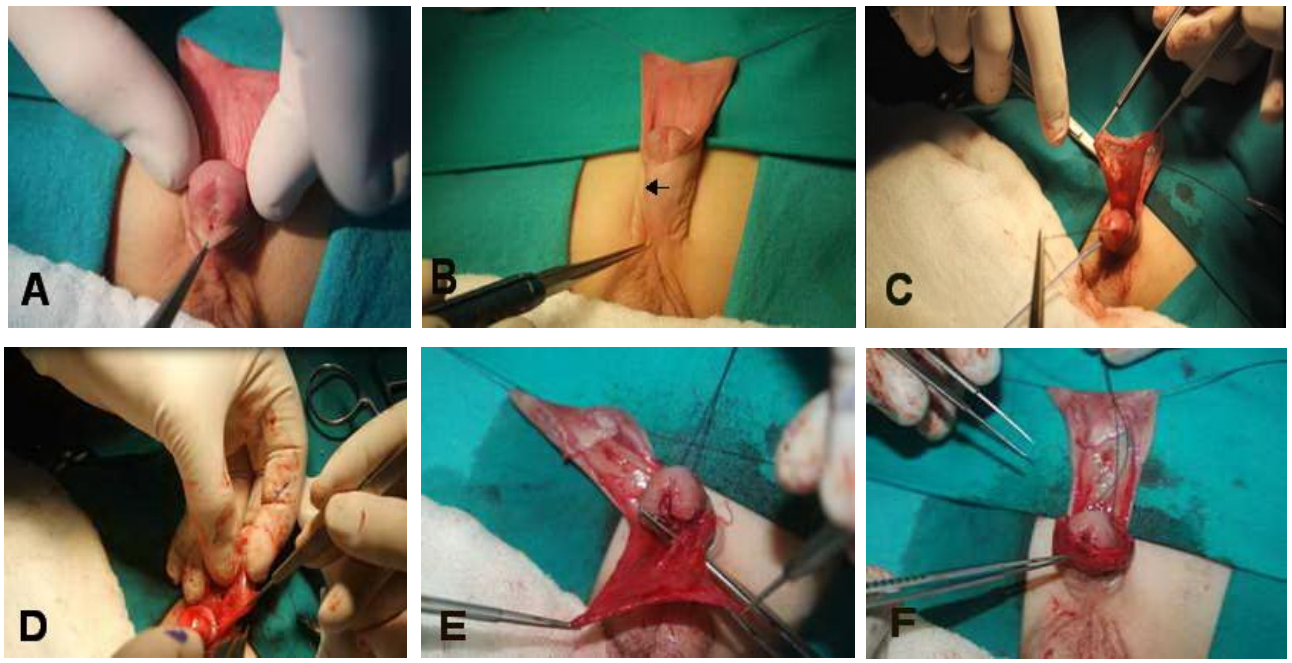

Fig. 1. A) Preop. Subcoronal hypospadias. **B)** Right deviation of penile raphe. **C)** Dorsal dartos fascial flap. **D)** Transposition of the Dartos fascial flap. **E)** Dorsal dartos fascial flap tumbling. **F)** Fascia flap supports new uretra.

Discussion

We encounter early and late complications after hypospadias surgical correction. As early complications, ischemia, hemorrhage, hematoma, infection and wound opening, and as late complications, most commonly urethral fistula, urethral stricture, urethral diverticulum can be seen. With the risk of low complication, cosmetic appearance and easy application method, the incision and tubularization concept has made the Snodgrass method popular in recent years, becoming hypospadias correction technique frequently used by surgeons. The most common complication after operation is fistula. Various methods have been described to reduce the possibility of fistulae [1-8,12,13]. Among the supports placed between the neo-urethra and the skin, the transfer of the dartos fascia is important. A longitudinal, well-vascularized dartos flap, harvested dorsally and buttonholed ventrally, represents a good choice for preventing fistula. [14]. In our case a proximally pedicled dartos fascia was formed and tumbled on penis to cover the entire neo urethra. The dartos fascia is an ideal choice for this purpose, with good blood supply and a very fine facial structure. While the blood of the fascia contributes positively to the healing of the region, it does not form a mass under the skin covered with both the upper and lower tissues. A solid barrier between the skin and the urethra provides aesthetic appearance.

Here, we will discuss two important issues in terms of the median raphe deviation: 1-median raphe deviation and hypospadias, 2-median raphe deviation and glanular torsion angle. It is known that the penoscrotal raphe deviation is rarely seen in infants. However, Mohan et al. [9] published evidence that penoscrotal raphe deviation occurs more frequently in association with certain conditions. They demonstrated that the approximately 88% of children with hypospadias had a median raphe deviation, whereas 86.3% of the children in the control group had a centrally positioned raphe [9]. In addition, the raphe deviation was mostly to the right (40.8%) in children with hypospadias and to the left in normal children [9]. In a study conducted by Sarkis et al. [11], 99% of patients with isolated penile torsion were shown to turn to the left.

There is a significant relationship between the median raphe position and the true angle of the glans torsion, but glans torsion was hidden in 4-12% of cases non- deviation [15-17]. By Pierrot et al [11], median raphe studied as a predictive factor for angle of glans torsion, and they suggested that the surgeon can predict with a simple external index (median raphe) the hidden angle of glans torsion before doing the circumcision and splitting the prepuce. Additionally, they demonstrated clearly that median raphe deviation at coronal sulcus and foreskin tip correlates to glans torsion.

Conclusions

As in our case here, a dorsal well-vascularized dartos flap that is buttonholed ventrally can be a good choice for the prevention of a fistula in the future in proximal cases [14,18]. In addition, evaluation of the penile median raphe is simple and can be a useful and reliable indicator of the presence of hypospadias in children with milder forms and glans torsion in the early period [5,11].

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: All photos were taken with parental consent.

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