INTRODUCTION

The true prevalence of congenital penile curvature (CPC) is difficult to determine. Some studies suggest that this problem may occur in as many as 10% of the male population [1]. However, a literature search of the Medline database revealed no reference concerning familial appearance of congenital penile curvature. For that reason we would like to present our case series.

MATERIAL AND METHODS

Two brothers aged 25 and 26 respectively were admitted to department of urology due to congenital penile curvature. Each patient was assessed by a history, physical examination, auto-photography of the erect penis, and a thorough sexual history. Concomitant anomalies of penile layers were absent in both cases.

A survey of the fetal penis at different stages of development shows some degree of curvature in a considerable number of embryos [4]. Penile curvature may thus be considered almost physiological in embryos between 35 and 45 mm in length. Thus, it has also been proposed that penile curvature is secondary to an arrest in normal penile development [5]. Therefore, some form of congenital local androgen deficiency may be responsible for inherited penile curvature.
(75° and 20°) curvature of the erect penis, respectively. Erectile function was normal. Both were qualified for surgical correction. In both cases, after degloving the penis with a tourniquet at the base, an artificial erection was created by injecting saline into the corpora cavernosa. (Figure 2)
A Yachia incisional corporoplasty [2] (Figure 3) was performed in the patient with ventral curvature (60°). Three pairs of longitudinal incisions of the tunica albuginea on the most convex side of the curvature in the dorsal part of the penis were made and then horizontally closed with continuous suture, Maxon 3.0.

In the second case (ventrolateral curvature), an Essed-Schroeder plicational corporoplasty [3] was introduced. Three pairs of reeving nonresorbable sutures were placed on the dorsal aspect of the tunica albuginea and one suture on the right hand side of the tunica albuginea to correct the curvature. Circumcisions were performed to minimize the postoperative complication rate.

The kinship of the patients was independently verified by analysis of short tandem repeats (STRs) commonly used in forensic genetics. A total of 15 autosomal and 18 Y-chromosomal STR loci were amplified by multiplex polymerase chain reaction and separated by capillary electrophoresis on a 3130 Genetic Analyzer (Applied Biosystems) (Tables 1, 2). Assuming prior probability of 50%, statistical analysis of the patients’ genotypes revealed posterior probability of 99.9999997% that they were full siblings, and thus confirmed their reported relationship.

RESULTS

Intraoperative check up revealed total straightening of the penis in both cases (Figure 4). The morbidity immediately after surgery was minimal. The patients’ satisfaction with the cosmetic and

| Locus     | BJ     | PJ     |
|-----------|--------|--------|
| AMELX/Y   | XY     | XY     |
| D7S820    | 9/10   | 9/10   |
| CSF1PO    | 11/11  | 11/11  |
| D3S1358   | 14/17  | 14/16  |
| TH01      | 6/9.3  | 6/7    |
| D13S317   | 8/11   | 8/12   |
| D16S539   | 12/12  | 11/12  |
| D2S1338   | 17/19  | 17/19  |
| D19S433   | 13/15  | 13/15  |
| VWA       | 15/17  | 14/17  |
| TPOX      | 8/8    | 8/8    |
| D18S51    | 15/16  | 14/15  |
| D5S818    | 10/12  | 12/12  |
| FGA       | 21/26  | 24/26  |
| D8S1179   | 13/14  | 12/14  |
| D21S11    | 29/29  | 29/29  |
A functional result was very good in long-term follow-up (Figure 5). There was no evidence of persisting numbness of the glans nor erectile dysfunction after treatment.

**CONCLUSIONS**

The true prevalence of congenital curvature is difficult to determine. Some reports suggest that this problem may occur in up to 10% of the male population, however, clinically significant bending is much less frequent [1]. The most common type is ventral, but several different varieties exist.

The precise etiology of congenital curvature of the penis is unknown. During normal embryologic development, the penis goes through stages of ventral curvature and then straightens. Penile curvature may thus be considered almost physiological in embryos between 35 and 45 mm in length. Kaplan and Lamm have shown that 44% of embryos 100-150 mm long still maintain some penile curvature, which may persist after the 3rd month of pregnancy [5]. For this reason, penile curvature is frequent in severely premature babies and may show spontaneous remission with growth.

Embryologic development of the penis is regulated by testosterone that is converted by 5-alpha-reductase to dihydrotestosterone (DHT). DHT binds to androgen receptors in all tissue layers in the penis. Catuogno et al. postulate that fetal androgen deficiency or local deficiency of 5-alfa reductase may be the cause of different penile malformations.

By the external application of a suspension of DHT (0.15 mg/cm²) once daily for 2-3 months, they have...
achieved almost complete straightening in five of 11 patients with penile deviations, but without simultaneous hypospadias and 30% improvement in two further patients [6]. Despite the fact that androgen deficiency may play a key role in the congenital penile malformations, we do not know what is the precise mechanism – environmental?, maternal?, fetal?, placental?

Histologically the deviation is associated with different abnormalities of the penile layers. Devine and Horton [7] proposed a classification system for congenital penile curvature based on that identifying three separate types of curvature. Type I is the most severe presentation. The structures normally surrounding the urethra (corpus spongiosum, Buck’s fascia, dartos) are absent. Type II presents normal differentiation of the corpus spongiosum, but with fibrous formation of Buck’s and Colle’s fascia. In type III congenital curvature, the patient’s urethra, corpus spongiosum, and Buck’s fascia are all developed normally, but the dartos fascia forms the elastic strip that causes the penis to bend. Kramer et al. [8] have proposed adding to the classification of Horton and Devine a fourth type, the curvature being due to asymmetry of the corpora cavernosa.

Microscopic study conducted by Darewicz et al. [9] revealed significant changes in the structure of the tunica albuginea such as chaotic alignment of the collagen fibers of different diameters, showing periodic widening and signs of disintegration and angulation.

In the two presented cases, the physical examinations showed penile curvature type IV according to extended Devine and Horton classification. Both patients were treated with standard procedures with good results. This is the first clinical report that attracts the attention of urologists to the familial form of penile deviation.

In our opinion further studies on CPC concerning epidemiology, genetics, histology, and embryology are needed. They may clarify if the CPC is a genetic disorder and what may be the best treatment.

References

1. Montag S, Palmer LS. Abnormalities of penile curvature: chordee and penile torsion. Scientific World Journal. 2011; 11: 1470-1478.
2. Yachia D. Modified corporoplasty for the treatment of penile curvature. J Urol. 1990; 143: 80-82.
3. Essed E, Schroeder FH. New surgical treatment for Peyronie disease. Urology. 1985; 25: 582-587.
4. G Marrocco, A Calisti, F Palmiotto, P De Simone, I Di Meo. Pediatr Surg Int. 1995; 10: 40-43.
5. Kaplan GW, Lamm DL. Embryogenesis of chordae. J Urol. 1975; 114: 769.
6. Catuogno C, Lanza T, Ventrice GA, Scalfari A, Lanza P. Medical therapy of congenital curving of the penis without hypospadias. Int J Impotence Res. 1994; 6 (Suppl 1): D225.
7. Devine CH, Horton CE. Chordae without hypospadias. J Urol. 1973; 110: 264-271.
8. Kramer SA, Aydin G, Kelalis PP. Chordae without hypospadias in children. J Urol. 1982; 128: 559-561.
9. Darewicz B, Kudelski J, Zynaka B, Nowak HF, Darewicz J. Ultrastructure of the tunica albuginea in congenital penile curvature. J Urol. 2001; 166: 1766-1768.