Management of perineal ectopic testes

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Prise en charge d'une ectopie testiculaire périnéale

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RÉSUMÉ

Prérequis : L'ectopie testiculaire périnéale est une malformation congénitale rare. Le testicule ectopique est situé entre le raphé penoscrotal et la région génitofémoral.

Objectifs : Nous rapportons 6 cas d'enfants traités pour ectopie testiculaire périnéale. Les aspects épidémiologique, clinique, radiologique et thérapeutique serons traités à la lumière des différents données de la littérature.

Résultats : entre 2000 et 2009 , 2156 malades ont été traités pour ectopie testiculaire au service de chirurgie pédiatrique à l'hôpital d'enfants de Tunis, six malades (0.2%) ont été traités pour ectopie testiculaire périnéale.L'âge moyen des malades était de 21 mois. Ces anomalies étaient associées à une hernie inguinale dans 2 cas. Le diagnostic était basé sur la présence d'une bourse vide avec une tuméfaction périnéale. Une orchidopexie in dartos a été, facilement, réalisée dans tous les cas par une voie d'abord inguinale. La longueur des vaisseaux testiculaires et du déférent étaient adéquates avec un trajet suffisant dans tous les cas.

Conclusion : La chirurgie en cas d'ectopie périnéale doit être réalisée dés la confirmation diagnostic. Elle est indiquée même en absence d'une hernie inguinale associée. Le diagnostic fonctionnel est difficile à définir mais semble identique à celui des autres sites d'ectopie testiculaire

Mots-clés

Ectopie testiculaire ; gubernaculum ; orchidopexie

SUMMARY

Background: Perineal ectopic testis (PET) is a rare congenital anomaly in which the testis is abnormally situated between the penoscrotal raphe and the genitofemoral fold.

Aim: we report six patients treated for PET. The epidemiological, clinical, radiological and therapeutic aspects of this rare entity are discussed in light of data of the literature.

Results: Between 2000 and 2009, six patients (0, 2%) treated for PET were diagnosed among 2156 patients operated upon for undescended testes in unity of paediatric surgery in Tunis children's hospital. The mean age was 21+/- 25 months. The abnormality was associated with an inguinal hernia in two cases. The diagnosis was based on the presence of an empty scrotum or perineal swelling. In all, orchidopexy in a dartos pouch was easily performed through an inguinal skin crease incision.The length of the testicular vessels and vas deferens was adequate with a favourable course in every case. Although the complications of undescended testes are the same as for PET, the timing of surgery should be different.

Conclusion: It is generally accepted that children must not be below 6 months of age for surgical correction of undescended testes, but there is no need to delay surgery in PET, which can easily be diagnosed by physical examination in the neonatal period. Surgery is indicated even if there is no hernia present. The functional prognosis, always difficult to define, appears to be identical to that of other sites.

Key-words

Cryptorchidism; ectopic testis; gubernaculums; orchidopexy

Testicular maldescence is the most common anomaly of the genitalia with incidence of 1.5%. It is reported that 5 % of maldescended testes are ectopic (1). Testicular ectopia specifically describes inguinoscrotal descent outside the normal boundaries. The five major sites of ectopic testes are the superficial inguinal pouch, femoral, suprapubic, transverse (contralateral hemiscrotum), and perineal. Perineal ectopic testis is a rare congenital anomaly in which the testis is abnormally situated between the penoscrotal raphe and the genitofemoral fold. Although the differential diagnosis between undescended (UDT) and perineal ectopic testes (PET) is easy and the complications are the same, the timing of surgery is controversial for PET and differs from that for UDT. In this report, we discuss the clinical features and appropriate time of surgery for PET.

PATIENTS AND METHODS

Between January 2000 and January 2009, six patients were treated for PET in unity of paediatric surgery in Tunis children's hospital. The mean age, side, associated anomalies, operation time, operation type, and results were evaluated.

RESULTS

The patients' mean age at admission was 21+/- 25 months (range: 2 months – 5 years). The follow-up time' mean was 4 years (1-9 years). One patient was admitted to the clinic with the diagnosis of strangulated inguinal hernia. The others were referred with the chief complaint of an empty scrotum which is associated in 2 cases to a perineal swelling (table 1). Two patients had an inguinal hernia on the same side. Right side was found in all cases. Physical examination revealed emptiness of scrotal content and a palpable ipsilateral perineal mass of normal shape and consistence (figure 1). There was no need for additional diagnostic examinations.

Figure 1 : Physical examination revealed emptiness of scrotal content and a palpable ipsilateral perineal mass.

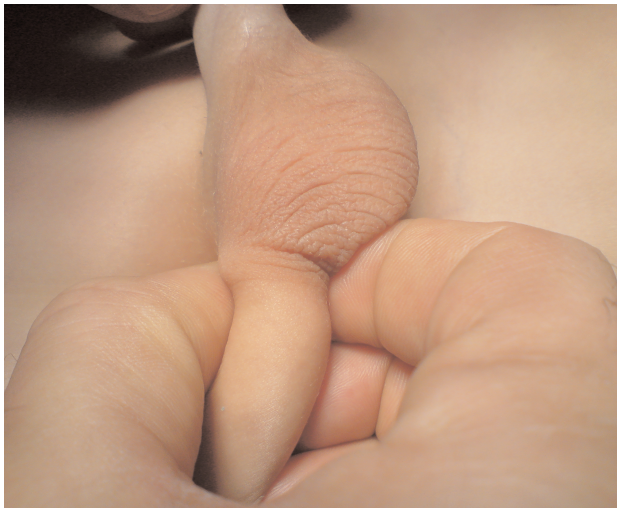

Table 1 : Patients data: R: Right; L: H: Hernia; S: strangulation

1	2 months	R	+H(S)	Perineal swelling
2	6 months	R		Perineal swelling
3	17 months	R		Empty scrotum
4	18 months	R	+H	Empty scrotum
5	2 years	R		Empty scrotum
6	5 years	R		Empty scrotum

All patients were treated surgically. Two patients were below 6 months of age at the time of operation. All patients were operated by a standard inguinal skin-crease incision.
During the surgical exploration, testis and guberbuculum were fixed to the perineum in all cases. ,The characteristics of the testicle and its elements were normal. Orchidopexy in the scrotal dartos pouch was performed easily in all cases. High ligation was also performed for two obvious hernias. Post operative recovery was uneventful.

DISCUSSION

PET is a rare congenital anomaly with incidence of 1% of all cases of undescended testis(2) since the first case of the disease was reported by John Hanter in 1786 more than 113 cases have been reported between 1786 and 1984 in English literature and 15 cases have been reported in Japanese literature at the same period (3).
Aetiopathogenesis of perineal ectopic testis is controversial. The role of the gubernaculum is not well elucidated. For some authors this ectopy results from a gubernaculum failure (4) and it's due to an anomaly of fixation of the distal extremity of the gubernaculum testis resulting in an abnormal position of the testis (5). The cremasteric muscles should be indicative of the course and the termination of the gubernaculums (6). Haston thinks that's due to an anomaly of the genitor femoral nerve which has a great role as a tractor and guide of the gubernaculum migration to the ectopic position (7). For other authors this ectopy testis may be due to a mechanic obstruction of scrotal inlet or in the entrance of the testis in the scrotum (which become fibrous) that leads the gonad to an abnormal position (4, 8).
Diagnosis is easily based on physical examination that reveals an empty, hypoplasic scrotum and a small mass lateral to it (4). Perineal ectopic testis is frequently associated an inguinal hernia and can sometimes be associated with other disorder such as hypospadias and scrotal anomalies (9, 10). A case of bilateral perineal ectopic testis was reported (11). The diagnosis can be made in antenatal diagnosis and may be performed by ultrasound at 38 weeks gestational age (2).
Echography can also study traumatic lesions and testicular vascularisation (5). Perineal testis can be discovered while a complication like torsion or hernia strangulation the same as undescendent testis (1).

Treatment is surgical and it's indicated as soon as the diagnosis is affected. There is no need to delay surgery in perineal ectopic testis so we haven't to wait for probable descent. Surgery is indicated even if there is no hernia present because of non negligible traumatisms and complications (1, 8). Testis is relocated and fixed into the scrotal dartos pouch through inguinal incision.

Orchidopexy is usually easily to perform so the length of the testicular vessels and vas deferens are adequate (12). Gubernaculum tests were usually found to lie fixed to the perineum and the ectopic testis and empty scrotum are often well developed (1, 13, 14). Perineal incision is not necessary and retroperitoneal dissection is not required while perineal adherences are frequent (6, 15). The long term prognosis seems to be excellent for some authors (1, 8) because of the histopathologic features involved, prognosis is better than that of cryptorchidism (12). In fact the natural history of the ectopic testis has been described as normal or less abnormal than that of a truly cryptorchid testis (13, 14). While Hatcheson, in a comparative study, has found similar pathological findings in ectopic and undescendend testis and suggests that ectopic and undescendend testis are variant of the same congenital anomaly (16). What's sure is that an early detection of the ectopy and its surgical repair on time favored the functionality of the testis.

CONCLUSION

Ectopic perineal testis is relatively rare but easily recognized by simple but attentive physical exam, so the less frequent localization of testicular ectopia should be palped. Surgical intervention should be early performed regardless of the existence of an inguinal hernia because there is no need to wait for probable descent. Orchidopexy by inguinal incision doesn't raise any particular problems because of the sufficient length of the spermatic cord. The functional prognosis difficult to define seems to be identical to that of other sites.

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