# An ovulating testis

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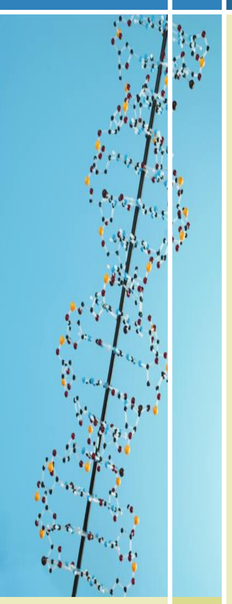
AUTHORS DISCLOSE NO CONFLICT OF INTEREST



**Background :** Ovotesticular disorders of sexual development (DSD) are a rare form of DSD with co-existence of both ovarian and testicular tissue in one or both gonads.

## Presentation

- A term infant (weight +1.38 SDS) presented at birth with severe penoscrotal hypospadias, a small phallus and a right hemiscrotum with descended gonad.
- External masculinization score was 1.5

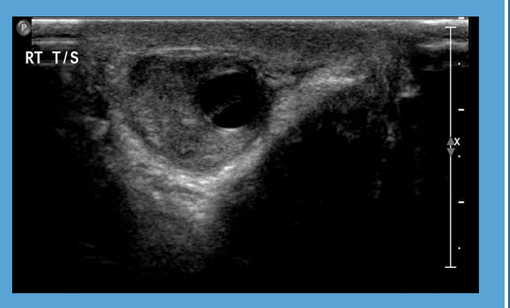
## Investigations

- Ultrasound revealed no mullerian structures, a small right gonad with probable epididymis, and no gonad on the left.
- Karyotype showed 46 XX with no mosaicism.

HCG test demonstrated functioning testicular tissue with a testosterone rise [7.9 to 3.4nmol/L]

| HCG Test | Day 3 | Day 6 | Day21 |
|---|---|---|---|
| Oestrogen | 361 | 165 | 71 |
| Testosterone | 7.9 | 13.4 | --- |
| 17 0HP | 179 | --- | 108 |

- Laparoscopy showed a vestigial uterus and a left gonad associated with fallopian tube which was removed.
- Histology confirmed ovotestes on both sides.
- Gonadal karyotype was 46XX.




- A diagnosis of 46XX ovotesticular DSD was made and a male gender was assigned with parental concurrence.
- He underwent hypospadias repair with good results.
- Family were keen to preserve gonad hence right ovotestis was left in the scrotum with a view to monitoring carefully at puberty.

## Subsequent presentation

- From age 13 there was evidence of virilisation.
- His testosterone was 4.4nmol/L, oestradiol 88nmol/l, LH 10.3 nmol/L and FSH 23nmol/L, indicating a failing gonad producing predominantly testosterone.
- Subsequently he developed progressive gynaecomastia.
- Repeat blood tests showed a fall in testosterone (0.8nmol/L) but detectable oestradiol (34nmol/L) levels.
- Hence a 3-day HCG test was undertaken (testosterone 1→14nmol/L; oestradiol 168→83nmol/L)
- 48 hours later he presented with acute right scrotal pain and underwent surgery.

## Progress

- Intra operatively he was found to be bleeding from the ovarian tissue within the testicular capsule.
- In view of progressive gynaecomastia and future malignant risk, his right ovo-testis was removed after extensive discussions with the family.
- Sperm counts prior to surgery had shown azoospermia and sperm harvesting was also unsuccessful.
- He had bilateral prosthesis sited and testosterone replacement commenced.

**Conclusion:** This case emphasises the complexity involved in the management of such rare conditions and the importance of systematic patient and family centred approach.