Bilateral inguinal hernia in a female child

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DESCRIPTION

A 15-month old phenotypic female child was brought to the clinic with fever and diarrhoea. A bilateral inguinal hernia with a solid palpable mass resembling gonads was observed ([figure 1](#)). There were no other symptoms and the remaining examination was unremarkable. An ultrasound evidenced absent uterus and ovaries and confirmed the presence of bilateral inguinal testis. The karyotypic analysis revealed a 46, XY karyotype.

Bilateral inguinal hernias are rare in female infants. Incidence of complete androgen insensitivity syndrome (CAIS) in these patients is 1–2%.^{1 2} The typical clinical presentation is either primary amenorrhoea in adolescence or inguinal swellings (later confirmed as testis) in a phenotypic female infant.¹

A patient with CAIS has the pubertal growth spurt at the appropriate age and oestrogen-dependent secondary sexual characters by aromatisation of androgens, with normal onset of breast development. The endocrine profile at puberty typically shows elevated serum levels of testosterone and luteinising hormone, due to the consequent lack of negative feedback.¹ Body proportions tend to be female or intermediate. There is an insufficiency of secondary sexual hair growth, with pubic hair growth usually not reaching Tanner stage 5.³

Figure 1 Bilateral inguinal hernia in a 15-month old phenotypic female child.

Management should be multidisciplinary and include gonadectomy to avoid malignant degeneration of the gonads held in the abdominal cavity, sex-hormone replacement and psychological support.¹ The real risk of cancer in CAIS gonads is low before puberty, so it seems safe to leave at least one gonad in situ until completion of spontaneous puberty.² Expected advantages are optimal development of breasts, improved bone density and preservation of self-esteem in an adolescent who will spontaneously achieve puberty.

Learning points

- ▶ Consider complete androgen insensitivity syndrome (CAIS) in phenotypic female children with bilateral inguinal hernia.
- ▶ Consider CAIS in girls presenting with an inguinal gonad, even if unilaterally.
- ▶ Management of CAIS patients should be multidisciplinary and include gonadectomy, sex-hormone replacement and psychological support.

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Competing interests None.

Patient consent Obtained.

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