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Surgery for intersex

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Abstract

Surgery for intersex is a complex and controversial topic. The traditional practice of surgery for all—usually during infancy—has been challenged by unsatisfied adult patients. There is at present little good outcome data partly due to the problems of non-disclosure of diagnosis, which leaves adult patients absent from follow-up studies. Only recently has more information become available on gynaecological and sexual outcomes. This review covers the three main aspects of intersex surgery: clitoral surgery, vaginoplasty and gonadectomy. Traditional and modern surgical techniques are described as well as any available follow-up data. The current controversies surrounding the role of intersex surgery are summarised.

Keywords: Intersex; Ambiguous genitalia; Vaginoplasty; Clitorectomy; Clitoral reduction; Gonadectomy

1. Introduction

The management of intersex conditions poses a huge challenge for the clinician. Almost every aspect of intersex is controversial but by far the most difficult and confusing area is that of surgery. There is no consensus as to whether or not surgery should be performed, especially for children with ambiguous genitalia at birth. If surgery is chosen, there is no consensus as to the most appropriate timing or which operation is best. Various new techniques and modifications of the classic techniques have been reported in the hope to minimise the morbidity that comes with surgery. However, there are few available studies on long-term outcome especially regarding psychosocial and psychosexual function in adult life.

This review will focus on the following three areas:

- Surgery for ambiguous genitalia.
- Vaginal reconstruction.
- Gonadectomy.

2. Surgery for ambiguous genitalia

Ambiguous genitalia are usually diagnosed at delivery when a newborn baby appears neither clearly male nor clearly female. Some of the conditions which present in this manner are given in Table 1. These babies are referred to a multidisciplinary intersex team comprising at the very least a paediatric endocrinologist, paediatric surgeon and psychologist. Complex investigations including genetic, hormonal and anatomical assessment should determine the underlying cause and assign a diagnosis. Once the diagnosis is reached, the child is assigned to the most appropriate sex of rearing. Sex assignment is a complex process that involves the parents as well as the multidisciplinary team. Each child should be reviewed on an individual basis and assigned to be raised as either male or female. Current standard practice in Europe and the USA is that those children assigned to the female sex of rearing will undergo feminising genital surgery. This practice is under review at present as whilst there is little dispute that each child needs to be raised either male or female there is considerable debate as to whether this allocation needs reinforcing with irreversible genital surgery.

Feminising genitoplasty comprises of surgery to the enlarged clitoris, vaginoplasty and labial refashioning.

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Table 1
Causes of ambiguous genitalia

46 XX karyotype
Congenital adrenal hyperplasia
21-Hydroxylase deficiency (most common)
11β-Hydroxylase deficiency
3β-Hydroxysteroid dehydrogenase deficiency
In vitro virilisation due to ingestion of virilising drug by the mother, e.g. Danazol
Placental aromatase deficiency
True hermaphroditism
46XY karyotype
Partial gonadal dysgenesis
Defect in testosterone biosynthesis
17β-Hydroxysteroid dehydrogenase deficiency
3β-Hydroxysteroid dehydrogenase deficiency
17α-Hydroxylase deficiency
True hermaphroditism
Androgen insensitivity syndrome—partial
Androgen receptor gene mutation
5α-Reductase deficiency
Leydig cell hypoplasia
Abnormal karyotype
Mosaicism and aneuploidy

3. Clitoral surgery

3.1. Techniques

There are three main types of surgical procedure for the enlarged clitoris: clitoridectomy, clitoral recession and clitoral reduction.

Clitoridectomy (or clitorectomy) was the first procedure performed to treat the enlarged clitoris. It involves amputation of the whole clitoris including the removal of its innervation and vascular supply. Clitorectomy is no longer practiced in the UK although is still the treatment of choice in some European centres [1]. As it is only recently that clitorectomy has fallen out of favour, many adult women will have undergone total clitoral removal as infants.

In order to preserve the innervation of the clitoris with the hope of minimising sexual dysfunction, various modifications to clitoral surgery have been reported. Clitoral recession involves burying the clitoral shaft under the pubic bone without removing any erectile tissue. This technique has become less popular due to reports of pain during erection especially when puberty is reached [2].

The third and now most commonly used technique is clitoral reduction. The glans clitoris and neurovascular bundle running along the dorsal aspect of the clitoral shaft are identified and preserved while removing most of the erectile tissue (Fig. 1). To further minimise the potential damage to the dorsal neurovascular bundle, other modifications to clitoral reduction have been designed. Ventral reduction clitoroplasty involves removal of erectile tissue from within the tunica via a ventral approach to reduce the size of the erectile body whilst preserving some erectile tissue and all the clitoral nerves.

Fig. 1. A clitoral reduction. The clitoral skin is reflected off the shaft of the clitoris. The neurovascular bundle on the dorsal aspect is isolated and preserved before removing the erectile tissue.

3.2. Outcomes

There is a paucity of good long-term follow-up data on patients following clitoral surgery. Studies are often small and anecdotal with no information on sexual function [3]. Well-organised adult patient peer support groups have raised awareness of potential difficulties as a result of genital surgery. This has triggered recent work on the effects of clitoral surgery on sexual function. The clitoris is an erotically important sensory organ and its only known function is in contributing to female orgasm. However, sexual response is multifactorial and the exact contribution of the clitoral glans, clitoral hood and clitoral corpora to orgasm is only poorly understood. Recent work on the anatomy of the clitoris has shown it to more extensive than previously thought extending behind the pubic bone to reach the anterior vagina [4]. Further examination of innervation of the clitoris has demonstrated nerves surrounding the tunica with multiple perforating branches entering the dorsal aspect of the corporeal body and glans [5]. Thus, any incision to the clitoral glans, corpora or hood may risk damage to the innervation. Past confidence amongst paediatric surgeons in unimpaired sexual function post clitoral surgery seems no longer tenable.

In a recent study, women with ambiguous genitals who had undergone feminising genital surgery were compared with women who had ambiguous genitalia without surgery [6]. Overall sexual function scores were poor in both groups when compared to a standard UK population of women. Those who had undergone clitoral surgery were significantly less likely to report experience of orgasm compared with