

# Sexual function and fertility issues in cases of exstrophy epispadias complex

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## ABSTRACT

In patients with EEC, the issues such as sexuality, sexual function and fertility gain more importance once these patients advance from puberty to adulthood. The aim of this review is to critically examine the available evidence on these issues. A systemic literature search was performed in Medline over the last 25 years using the key words: Exstrophy, sexual function and pregnancy. Search results were limited to studies of patients with exstrophy published in English literature. A total of 1500 publications were found and subsequently screened by title and when appropriate by abstracts. Of these, 40 publications pertinent to the subject were included for the analysis. The publications were supplemented by an additional 15 publications obtained from their bibliographies. The studies were rated according to the guidelines published by the US department of health and human services. Heterosexuality is usually expressed in both the sexes and most of them have adequate sexual function. Urinary diversion in some series seems to result in better ejaculatory hence fertility outcome in male patients. Recent series have shown equally good results with primary reconstruction. Most of the female patients have normal fertility while male patients have significantly low fertility. Most of the male and female patients with EEC have adequate sexual function. Most of the female patients have normal fertility while most of the male patients have significantly low fertility.

**Key words:** Exstrophy, epispadias, fertility, pregnancy, sexuality, sexual function

## INTRODUCTION

Over the last three decades, the refinements in the techniques of exstrophy repair have resulted in tremendous improvement in functional and esthetic outcome in cases of exstrophy epispadias complex (EEC).<sup>[1-4]</sup> Today, we see more and more adolescent and adult patients who not only have good urinary continence but also have well preserved upper tracts. These patients have the same life span as any healthy individual in a modern society. Like any one, they have their own dreams and ambitions, to lead a

normal social life and nurture the desire to have children and a family of their own. For them, sexual function and potential to produce a child become more important issues once these patients advance to adulthood. Many authors have tried to address these issues but barring few most of the series have small number of patients with limited information and follow-up.<sup>[1-4]</sup>

## DISCUSSION

Heterosexuality is usually expressed in both males and the females with EEC. Still a small number of male patients (10-20%) may not show any established relationship with women.<sup>[5]</sup> The beginning of sexual activity is usually delayed. Dissatisfaction with genital appearance and size was associated with emotional and body image disturbances, more commonly seen in males (30-60%).<sup>[5]</sup> Nearly three fourth of both male and female patients experience normal orgasm with partners' satisfaction. Both the sexes have shown a feeling of insecurity and worry about their sexual relationships and its long-term stability.<sup>[5]</sup>

Although the penile length is short, most male patients are reported to have normal libido and erections adequate for sexual intercourse.<sup>[4,5]</sup> Magnetic resonance imaging (MRI)

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studies have shown that short penile length is secondary to congenital shortening of anterior corpora rather than the pubic diastasis. The total corporal length was 60% greater in controls (16.1 cm versus 10.1 cm in EEC). This deficiency was mainly evident in anterior corpora.<sup>[6]</sup> Therefore, in spite of adequate closure of pubic symphysis diastasis, the penile length is shorter in EEC cases as compared to the normal individuals. The diastasis, over the time is known to recur with varying degree after repair, is not associated with penile shortening. Short penile length, more commonly than penile deviation and penile curvature (34-45%), is reported to be distressing (2-35%) in many of these patients.<sup>[6,7]</sup>

The prostate gland, an important component of male reproductive system, is unique in its configuration. It is located noncircumferentially around the urethra and later being anterior to the prostate gland. The volume and histology of prostate gland are comparable to normal men. The seminal vesicles are normally present and can be distended secondary to stasis and impaired drainage from previous surgeries.<sup>[6]</sup>

A significant number of patients do not ejaculate normally and suffer from varying degree of seminal fluid abnormality. Normal sperm count has been reported in 16-63% of these patients.<sup>[8,9]</sup> Although, in various series, 35 to 93% of the patients are reported to have ejaculation but the volume of ejaculate is low (mean <1.6; range 0.2-7.0 ml).<sup>[1,4,7,9]</sup> Nearly 50% of all these patients experience retrograde ejaculation. Ejaculatory or paternity problems have more commonly been reported in patients who have undergone genital reconstruction or cystectomy as compared to those who underwent early urinary diversion.<sup>[1,3,4,8]</sup> Azoospermia was reported in nearly 50-75% in such cases. Stein *et al.*, reported that five cases who did not undergo genital reconstruction had normal ejaculation, while 83% of the 28 patients who underwent genital reconstruction had post orgasmic semen dribble only and remaining 17% had no ejaculation at all.<sup>[4]</sup> A review by Woodhouse showed that the paternity was better preserved (almost twice) in patients who had undergone urinary diversions as compared to those who underwent genital reconstruction.<sup>[8]</sup> Contrary to this, Ben-Chaim *et al.*, reported 12 (75%) of the 16 patients experienced normal ejaculation and orgasm. Fourteen of these 16 patients had under gone staged primary reconstruction.<sup>[1]</sup> More recently, Anne *et al.*, reported on 21 patients who underwent single stage repair, 19 (90.50%) had ejaculation. The authors concluded that early genital surgery and functional bladder neck reconstruction did not result in adverse ejaculatory and fertility outcome. Infact, functional bladder neck reconstruction with anatomical placement of the colliculus seminalis in the posterior urethra allowed antegrade ejaculations in over 90% of the patients.<sup>[9]</sup> Thus, it may be inferred that nearly three fourth of the patients in both the groups (diversion and genital reconstruction)

have some form of ejaculation, only the force varies.<sup>[1,3,4,8,9]</sup> Further, the better ejaculatory outcome in the last two series with sizable number of patients leaves this question unanswered whether primary reconstruction really affects these functions adversely?<sup>[1,9]</sup>

It is difficult to exactly find out the fertility rate in these patients as in most of the series the age of the patients is comparatively young and follow-up is short. Besides, it is difficult to find out how many patients in actuality desired to produce children. It was interesting to note that in a review that nearly half of the patients who actually desired to have children were ultimately successful.<sup>[3,8]</sup> The various causes contributing to low fertility (average 15%) in these patients described are: Reconstructive surgeries leading to vassal damage, recurrent infections (epididymitis; nearly one third of the patients) and retrograde ejaculation secondary to iatrogenic injury to the bulbospongiosus muscles and the absence of circular prostate that contributes to normal bladder neck closure under sympathetic stimuli at the time ejaculation.<sup>[1,3,4]</sup> Such a low rate of fertility in male certainly raises the question whether in these patients the surgery is performed at the cost of fertility? However, one should weigh the benefits of reconstruction to achieve the functional and esthetic outcome in these patients, which remain the main objectives until puberty. Today, we have assisted reproductive techniques available to help these patients to father their own children. Testicular sperm extraction and intracytoplasmic sperm injection (ICSI) has been of great help in many of these patients.<sup>[3]</sup>

Most of the females with EEC have normal sexual desire and are adequately sexually active. Menstruations are mostly normal. Like penis in males, the vagina in females is short and vertically lying, which may need introitoplasty or vaginoplasty for normal sexual intercourse.<sup>[1,4]</sup> The associated problems which may adversely affect the sexual activity are: Leakage of urine during sexual intercourse, reduction or lack of clitoral sensation, dyparunia (25-50%) and genital prolapse (30-60%).<sup>[1,10]</sup> The latter is the most common and troublesome complication and tends to get precipitated during pregnancy. That is why many authors recommended an elective suspension procedure.<sup>[4]</sup> Uterine prolapse which is recurrent or non responsive to pessary insertion may need osteotomy and re-adaptation of pelvic floor muscles as to provide support to uterovaginal unit. Genital reconstruction in females is not that complex as in cases of males; hence, these patients have better psychosexual outcome.<sup>[1,5]</sup> In contrast to the males, most of the females have normal fertility. The fertility problems reported in some of these patients may would have been secondary to extensive pelvic dissection and tubal injuries.<sup>[1,3,8]</sup> The first case of pregnancy in women was reported by Bonnet in the year 1724 and since then many other authors have reported similar incidents without any adverse fetal outcome.<sup>[10]</sup> Shapiro *et al.*, reported 164

pregnancies in 131 women and more recently Mathews *et al.*, reported 11 pregnancies in 83 such patients.<sup>[1,10]</sup> All women with exstrophy should be adequately counseled during pregnancy and may be advised early bed rest in second trimester to reduce the likelihood of premature labor. Although vaginal deliveries are reported, it is mandatory to have elective caesarean section as to avoid any risk of injury to the reconstructed urinary tract, which might jeopardize continence.<sup>[2,3]</sup>

Finally, the data on hormonal profile involved in reproductive physiology in patients with EEC are as such sparse. The hormonal status is usually normal barring minor variations in some reports.<sup>[9]</sup>

## CONCLUSIONS

Most of the male and female patients with EEC have adequate sexual function. Nearly three fourth of the male patients are capable of ejaculating with varying force. Urinary diversion in some series seems to result in better ejaculatory hence fertility outcome. Recent series have shown equally good results with complete reconstruction. Male exstrophy patients are reported to have more of psychosexual disturbances as compared to the females. Most of the female patients have normal fertility but contrary to this most of their counterpart male patients have significantly suboptimal fertility.

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