The short-term disadvantageous of tension-suture and spica cast application in exstrophy-epispadias complex reconstruction: a case report

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Abstract

Background: Exstrophy-epispadias complex (EEC) represents a spectrum of genitourinary malformations and associated with another congenital anomaly. EEC is a rare congenital disability. EEC affects the genitourinary system, anterior abdominal wall, musculoskeletal system, spine and anus. The surgical outcome has a lot of potential complications in patient with EEC. The decision to first approach is important for the next step. Herein, we reported a case of EEC with several anomalies.

Case Description: A new-born male was born with section cesarean at 38 weeks of gestation from 34-year-old mother, and this is from the second pregnancy. The weight was 3300 grams, and the height was 48 centimetres. There was an abdominal wall defect with an exposed evaginated bladder plate with urine draining from the defect. A complete dorsally opened urethral plate was seen from the bladder neck down to the glandular groove. The patient was diagnosed with Exstrophy-epispadias complex (EEC). A 2 stage repair was conducted in this patient. After 10 days post-operation, the wound became dehiscence and got sepsis postoperatively as well as lead to die in the day 20.

Conclusion: Wound dehiscence following reconstruction is very critical inpatient with EEC. Tension suture and spica cast application can be disadvantageous inpatient with EEC.

Keywords: exstrophy epispadias complex, congenital malformation, pediatric, reconstructive.


INTRODUCTION

Exstrophy-epispadias complex (EEC) is a congenital anomaly of the genitourinary system with congenital malformations ranging from isolated epispadias to classical bladder exstrophy, and cloacal exstrophy is the most severe presentation.1 Besides, EEC also involved musculoskeletal system, pelvic floor musculature, and bony pelvis anomalies.1 Bladder exstrophy is a rare congenital malformation, with an estimated incidence of approximately 1 per 50,000 live births and affecting males approximately twice as often as females.2 EEC is caused by a derangement in mesodermal layers fusion during the first weeks of fetal life.3 At the end third week of gestation, intermediate layer of mesoderm starts to invaginate to give origin to the urogenital system, while the lateral plate mesoderm will contribute to forming the primitive gut tube.3 A disruption in this interaction is related to a cloacal membrane overgrowth and is prevent medial migration of mesenchymal tissue. EEC can be diagnosed prenatally with fetal transabdominal ultrasound (US) between the 15th and 32nd weeks of pregnancy.3,4

Several studies have been concerned about the surgical treatment need to be provided among patients with EEC. A surgical closure approach on bladder exstrophy in the neonatal period is the current primary method of care for most infants.4 A majority of these patients were known undergone either complete primary closure or modern staged reconstruction.4 The success of initial bladder exstrophy closure is essential for the development of adequate bladder capacity and future urinary continence.4 In order to reach this goal, a tension-suture and spica cast application techniques have been mentioned as one of appropriate interventions among EEC patients.5,6

A tension-suture method has been preferred for patients who undergone abdominoplasty procedures.7 This suture-technique is known can reduce the local complications, including hematoma and seroma formation, flap necrosis, and hypertrophic scars in this procedure.7 This technique also could be implemented on EEC patients. Besides, a conservative approach by increasing the immobility post-operatively is required to reduce the complications. Spica-cast application has been well known for its safety and efficacy for immobilization following initial bladder closure in classic bladder exstrophy.7 However, the short-term disadvantageous of
tension-suture and spica cast application in exstrophy-epispadias complex reconstruction has not been well documented. Based on those mentioned above, this study aims to evaluate the case report in showing the pitfall following EEC operation.

**CASE REPORT**

A new-born male was born with section cesarean at 38 weeks of gestation from 34-year-old mother and this is from the second pregnancy. The weight was 3300 gram, and the height was 48 centimetres. Previously, the mother has had a spontaneous abortion on first pregnancy at 28 weeks' gestation with unknown aetiology.

The husband was 36-year-old — the new-born presented with a genitourinary defect and polydactyly on right thumb (Figures 1A and 1B). There was an abdominal wall defect with an exposed evaginated bladder plate with urine draining from the defect, a complete dorsally opened urethral plate was seen from the bladder neck down to the glandular groove, both corpora cavernosa were clearly visible beneath and alongside the urethral plate, the scrotum was normal with descended testis on both side. There was no associated anorectal malformation (ARM), the anus was present, baby had meconium when he was born and was able to defecate. Echocardiography examination found ASD and PDA. The PDA defect closed spontaneously after 46 days and right after that, we decided to perform two stages reconstruction for this patient. We also found indirect right inguinal hernia. These genitourinary anomalies were consistent with EEC. Chromosome examination was held and revealed 46XY. Haematology examinations including full blood count, urea, creatinine and serum electrolytes were performed and the results were normal. Babygram radiology showed separation of pubic symphysis more than 4 cm and external rotation of anterior segment pelvic bone more than 18°. Intertirradiate distance also increases (Figure 1C). Delayed closure in this patient leads to further inflammatory or mechanical alterations with signs of mucosal inflammation such as a whitish coating (Figure 2A). We were planning to perform 2 stage repairs in this patient. The first stage was osteotomy by orthopaedics surgeon and bladder reconstruction (Figure 2B, 2C and 2D). We performed bladder neck and epispadias reconstruction on the second stage. However, after 10 days post-operation, the wound became dehiscence (Figure 3) and patient has sepsis due to the dehiscence. Eventually, at 20 days, a post-operation patient died because of sepsis condition.

**DISCUSSION**

In most EEC cases, the bladder and abdominal wall should be closed soon after birth. The standard-staged repair, primary closure of the bladder without osteotomy, and genital reconstruction must attempted in the first 72 hours of life to performed reconstruction. At birth, hamartomatous polyps were seen on the exstrophy bladder surface in about 50% of the cases. These polyps have been interpreted as reactive, potential pre-malignant environmental changes. Closure of the bladder within the first few hours of life is widely recommended in-patient with exstrophy bladder. In this case, we performed bladder reconstruction after the PDA closed spontaneously on 46 days.

Indirect inguinal hernias inpatient with EEC is due to a persistent processus vaginalis, large
CONFLICT OF INTEREST

There is no competing interest regarding the manuscript.

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AUTHOR CONTRIBUTION

All of the authors are equally contributed to the case report from data gathering until reporting the outcome of study.

REFERENCES


Figure 3  Wound dehiscence in 10 days post-operative