Primary Repair of Cloacal Exstrophy with a Small Bladder with a Bowel Patch: A Case Report

Johann Paulo S. Guzman[1,2], Ivory June J. Cadete[2], Ryan Ed S. Comuelo[2], Demie O. Tidon[2]

[1] West Visayas State University, College of Medicine, Department of Human Biology, Iloilo City, Philippines
[2]Western Visayas Medical Center, Department of Surgery, Iloilo City, Philippines
Corresponding author: jpguzman@wvsu.edu.ph

Abstract

Cloacal exstrophy (CE) is a group of congenital anomalies most commonly affecting the genitourinary, gastrointestinal and skeletal systems. Management of these anomalies requires multidisciplinary care and involves staged surgical reconstruction. We present the case of a newborn male referred due to a midline abdominal mass and imperforate anus noted upon delivery. Physical exam revealed an omphalocele, bladder exstrophy, bifid penis, and imperforate anus. Imaging revealed right renal agenesis and pubic diastasis. The patient underwent repair of cloacal exstrophy, bilateral ureteral stent, bilateral anterior osteotomy, primary repair of bladder with bowel augmentation to increase capacity, perineal urethrostomy, genital reconstruction and end colostomy. He was maintained in a pelvic wrap for the osteotomy to heal. He was discharged well. The surgical management of exstrophy is varied involving multiple approaches. While most authors would do a staged repair of the exstrophy leaving the bladder augmentation later because of high failure rates. Our approach to primarily address a relatively small bladder with a patch during closure in selected patients is feasible. Although long term follow-up is needed to achieve good quality of fecal and urinary incontinence, early surgical bladder correction may suggest earlier achievement of continence among patients. Patient is doing well after three years of this procedure with minimal soiling per urethrostomy. Our future plans include repair of the imperforate anus and urodynamic studies to monitor bladder function.

Keywords: extrophy, imperforate anus, urinary incontinence, bladder dysfunction, osteotomy, newborn

ISSN 2244 - 4335 (print)
ISSN 2651 - 6659 (online)
https://doi.org/10.59460/wvsurjvol12iss2pp1-12

INTRODUCTION

Extrophy belongs to a constellation of anomalies; OEIS complex (Omphalocele, Exstrophy of the Cloaca, Imperforate Anus and Spinal Defects) (Sadler, 2019; Cervellione et al., 2015). It is in close relation within the spectrum of the exstrophy–epispadias complex (EEC) of congenital anomalies. It is estimated to occur in about 1 in 200,000 to 400,000 births worldwide. (Sadler, 2019) Other anomalies in this spectrum include persistent ductus arteriosum, intestinal malrotation, renal agenesis, and congenital hip dislocation. In a survey of major European centers (excluding Norway and Ireland) treating congenital anomalies over a 12 - month period, they identified a total of 238 babies with exstrophies, 21 (8.8%) of them have cloacal exstrophies. Most of them are males (n=17, 80.9%) (Cervellione et al., 2015). In this same report, they noted 71.4% incidence of associated anomalies in cloacal exstrophies compared to just only 2.8% incidence among patients with only epispadias. These associated anomalies were described as cardiac, anorectal, orthopedic, central nervous system problems and trisomy 21 (Cervellione et al., 2015). These anomalies are often difficult to manage with involvement of multiple subspecialties (Sadler, 2019; Musleh et al., 2022). While it is established that cloacal exstrophies can have a myriad of other issues in the acute setting, a recent systematic review involving 12 studies have shown that chronic problems can become persistent. This includes urinary incontinence ranging from 9.1-85%, sexual function issues related to vaginal anomalies (8.3 – 71.3%) or uterine anomalies (14.3—71%), gender identity issues in 46 XY patients raised as females (11.1 – 66.7%), reduced ambulatory capacity (13.8%) and with only one paper reporting depression among gender reassigned patients (Musleh et al., 2022). Although, there is a wide range of reported values in that study, this could be probably due to the variation of definition used in the studies included in the review. Nonetheless, this provides a description of anomalies associated in both the acute and long term setting in the management of these patients.

Locally, the University of the Philippines- Philippine General Hospital reported 23 cases from 1983 to 2002 in one published case report of delayed repair in a bladder extrophy (Magahin & Alviar, 2010). After this, there was no recent updated report on their census nor have other centers in the Philippines reported their experience. In our center, for the past years, we had cases of abdominal wall defects. We had two cases of exstrophies in the last five years. We report our experience of a cloacal exstrophy in which we decided to repair it primarily with a patch of bowel to achieve tension free closure.

Authors of reported studies have recommended staged closures and pelvic osteotomies for a successful closure of an exstrophy as they have recognized these are independent factors for higher success rates of closure (Jayman et al., 2019; Sholklapper et al., 2022). In a multivariable analysis, statistically significant Odds Ratio (OR) were reported among patients using staged repairs compared to single stage [OR 3.7 (1.2-1.5)] and those who had osteotomies [OR 5.8 (1.7-19.60)]. Interestingly, they have also reported statistically significant higher closures if the osteotomies were done as staged procedure and prolonged immobilization (4 weeks) after closure (Jayman et al., 2019).

We argue that having staged procedures are costly and these are not usually reported in most studies. Thus, we have developed this technique to try to lessen the number of operations a patient will need to undergo for correction. From a technical standpoint, osteotomies are fundamentally needed because they lessen the tension during closure and can increase space in the pelvic cavity for the bladder. This report highlights that cloacal exstrophies with a difficult to close small bladder can be successfully done with a bowel patch augmentation in carefully selected patients. Our approach maybe comparable to contemporary staged approaches as we gain experience in treating more patients.

Case Report

This patient was delivered term by a 29-year-old G2P2 (2002) mother via normal spontaneous delivery and had an unremarkable perinatal course. Regular prenatal check-ups were initiated at the 3rd week of pregnancy in accordance to contemporary obstetric guidelines. There no recalled history of antenatal medication intake, illness or radiation exposure. Maternal and paternal family history were unremarkable. Maternal past medical history was also unremarkable. A screening ultrasound done during the second trimester reportedly revealed no congenital anomalies. At birth, there was note of a midline abdominal mass and the absence of an anal opening. Patient was coordinated to our institution at the second day of life for subsequent management. He came in our institution active with good activity and good suck. Physical examination showed symmetric chest expansion, an adynamic precordium, no murmurs and clear breath sounds. He had a globular abdomen with 3 cm omphalocele extending to the inferior abdomen. The bladder plate measuring 3 x 2.5 cm was exposed and flanked by two hemiphalluses. He had no anal opening and had non-palpable testes in the scrotum (see Figure I).

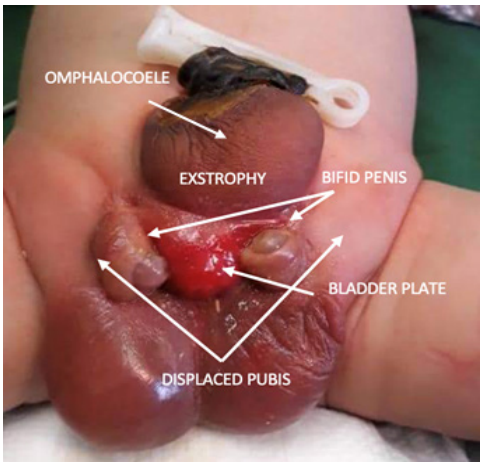

Figure 1. Cloacal exstrophy, bifid penis, small bladder plate, omphalocele and a widely displaced pubis symphysis

Laboratory results revealed anemia, which was corrected pre-operatively. His hemoglobin level rose from 11 g/dL on admission to 18 g/dL pre-operatively. Serum bilirubin levels (>1.2 mg/dl) and C-Reactive Protein (1.0 mg/dl) were elevated. Other laboratory findings were within normal limits. Chest x-ray revealed bilateral pneumonia. Whole abdominal ultrasound, to look for other associated anomalies, revealed an infraumbilical anterior abdominal wall defect with protruding masses exhibiting peristalsis. The urinary bladder and right kidney were unidentifiable. The liver, biliary tree, spleen and left kidney were sonographically normal. (Figure II). Echocardiography and spinal ultrasound were unremarkable. Spinal radiographs were normal (Figure III).

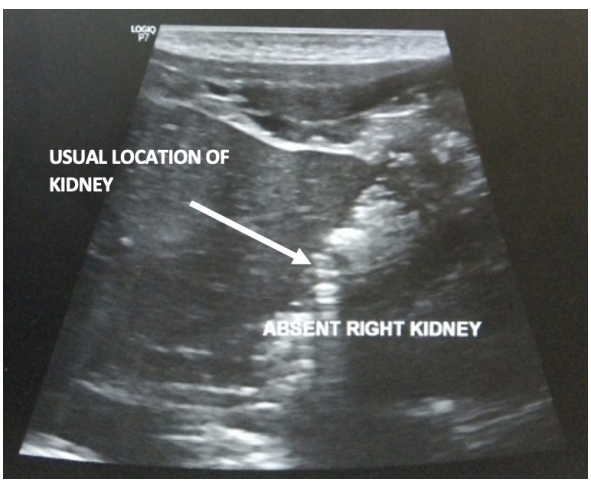

Figure 2. Ultrasonography showing absent right kidney

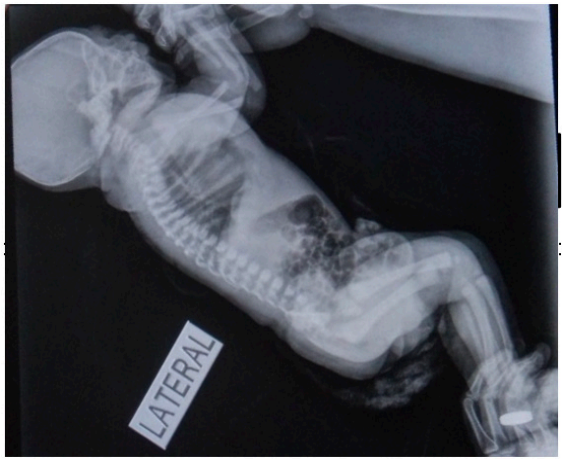

Figure 3. Lateral x-ray showing a grossly normal spine, no defects.

He underwent surgery on the fourth day of life. Intra-operatively we noted: 1) very small bladder capacity, 2) distal sigmoid attached to the posterior bladder wall with common opening of both the ureter and the hindgut, 3) pubis displaced 3 cm apart; 4) omphalocele size of 3 cm, and 5) absent urethral opening. We repaired the cloacal exstrophy and inserted bilateral ureteral stents for urinary diversion. We opted to do bilateral anterior osteotomy so that we will not convert to a prone position for a posterior osteotomy. The bladder was fully mobilized until a tension-free primary repair can be done. The urethral opening was tubularized and placed in the perineum, essentially creating a perineal urethrostomy. Genital reconstruction was done with re-approximation of the two hemi-phalluses. The intestinal segment was separated from the posterior bladder and the distal bowel was fashioned as an end colostomy. We incorporated 3 cm x 2 cm of hindgut intestinal segment to the bladder repair because the native bladder (1.5 cm x 1.5cm) was relatively small. The incorporated bowel allowed us to repair the bladder without tension. The bladder capacity was also increased after the incorporation (measuring 3.5 cm x 3 cm). The omphalocele sac was excised. The abdominal wall and pelvic floor were approximated (Figure IV).

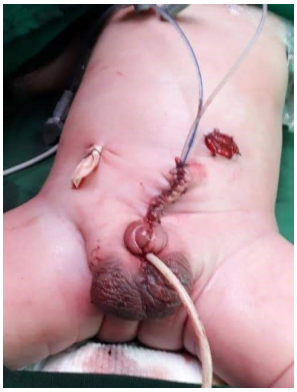

Figure 4. Patient after the cloacal exstrophy, genital reconstruction, end colostomy, closure of bladder with urethral stent and ureteral stents.

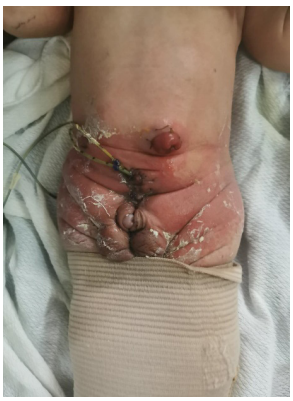

Figure 5. Patient maintained in a pelvic wrap

The patient was transferred to the Surgical Intensive Care Unit and was extubated on the first post-operative day. Feeding was eventually started on the 9th post-operative day due to ileus. The patient was maintained in pelvic wrap for four weeks to stabilize the anterior osteotomy (Figure V). He developed sepsis with Methicillin Resistant Staphylococcal Aureus (MRSA) infection on the 12th post-operatively day and was given vancomycin. The rest of the post-operative course (12th day up until the day of discharge) were unremarkable with good weight gain, normal urinary output (1-2ml/kg/hr) and mushy stools per ileostomy. Bilirubin, white blood counts and C-reactive protein levels normalized at the 15th hospital day. He was discharged well after completion of antibiotics with good activity on the 21st hospital day. The wrap was removed after one week of follow-up with no surgical site infections. He is on regular follow –up every six months with us for three years now with good weight gain (adequate Z-scores for weight and height), no difficulty in ambulation and minimal soiling per urethrostomy.

Ethical Considerations

This case was under the service of the Department of Surgery Western Visayas Medical Center in collaboration with some faculty members of the West Visayas State University College of Medicine. Informed consent was taken from the parents for the publication of this report and recorded. In line with the Data Privacy Act of 2012, no identifying patient markers such as hospital number, exact dates of admission and address were included in any part of writing of the manuscript process. This is not eligible for the ethical board review board of the hospital since this is already post-hoc. Patient and guardian's anonymity were insured. This case has been discussed and approved by the Technical Review Research Committee, Department of Surgery of the Western Visayas Medical Center.

DISCUSSION

CE is often manifested by a classic set of anomalies that include: exstrophy of the bladder and terminal ileum, complete phallic separation, pubic diastasis, a rudimentary hindgut, imperforate anus, and an omphalocele (Morales et al., 2016). The exact pathogenesis of this group of anomalies remains unknown. Theories include a) failure of mesenchymal cell migration during the 4th week of gestation b) disruption of the intermediate layer of the mesoderm in the 5th week of gestation and c) premature rupture of the cloacal membrane (Gearhart & Di Carlo, 2020). While being part of a wider spectrum of congenital anomalies, CE also presents with a spectrum of systemic involvement, most often presenting with anomalies of the genitourinary, gastrointestinal, and skeletal systems. Thus most cases, including ours, the long term survival and outcomes is often dictated with the severity of associated anomalies. Almost half of patients with CE have renal anomalies, the most common of which is a solitary kidney. It is 30 times more likely to occur in patients with CE than in those with bladder exstrophy alone. Males with CE often have cryptorchidism and separation of the penis into two hemiphalluses. Ninety percent of the time, patients have an omphalocele. Other associated intestinal abnormalities include imperforate anus, malrotation, intestinal atresia, ileal intussusception, and Meckel diverticulum. Sixteen percent of patients with CE have hip dysplasia. In our experience it is advisable to be meticulous during surgery as to not to miss out on these things. We advise immediate correction of identified anomalies together with the exstrophy if technically feasible. If a stoma is needed, we advise to place it away from surgical sites as far as possible to avoid infections similar to what is shown in Figure V. The most common spinal cord anomaly associated with CE is

dysraphism of the lower spinal cord, which occurs in up to 90% of patients (Gearhart & Di Carlo, 2020; Weiss et al, 2020) This contributes to urinary and fecal incontinence. We also recommend that initial work-up including spinal radiographs should be taken prior to correction to assess continence prognosis.

After birth, the exposed structures should be protected using plastic wrap. Hydration and nutrition should be optimized as soon as possible. A multi-disciplinary approach, involving pediatrics, pediatric surgery, urology, and plastic surgery is ideal. Since CE is part of a spectrum, the operative management of each patient is unique. The principles, however, include: 1) closure of the omphalocele, 2) reapproximation of the posterior part of the bladder, 3) end colostomy, 4) repair of the exstrophic bladder and genitalia, and 5) approximation of the pubic diastasis. Surgery often requires a multistage reconstruction as what others have suggested (Musleh at al., 2022; Jayman et al., 2019 ;Gearhart & Di Carlo, 2020; Weiss et al, 2020) In this case, bladder reconstruction was done with a single-stage approach. We incorporated part of the bowel wall on the repair and we acknowledged that this might potentially lead to a formation of a diverticula as the child grows. We opted to do this and monitor this patient for that possibility in the future. Without incorporating that small patch of bowel, a tension-free closure might not be feasible. This also led to an increase in bladder capacity. Some other centers have approached with repairing exstrophies in a single stage. In one Indian Urologic Center, out of 15 patients operated for CE, 5 (33.3%) died post-operatively due to prematurity and low birth weight. They had one bladder dehiscence (Dutta, 2014). It was not clear whether they included bowel augmentation or they simply reattach the hemi bladders in their closure. Similar to our stand, they also suggest that primary closures are more feasible to term babies with adequate weight.

In the past, bladder exstrophy was managed with cystectomy, however, patients often died due to renal complications (Inouye et al., 2014). Current surgical approaches allow bladder preservation and continence. The bladder may undergo normal development and obtain sufficient capacity after successful closure (Mathews et al., 1999) However, in some cases, the bladder may remain small, or the initial primary closure may fail. Others have reported some success in using porcine small intestinal mucosa or cadaveric skin graft for primary closures in some patients with small bladders. This has yet to be widely accepted probably due to the availability (Admire, 2013). Usually, our approach is that if the bladder template is small and technically difficult, reconstruction may be delayed. In patients who underwent closure but do not develop sufficient bladder capacity, augmentation cystoplasty may be indicated in the future. Thus we request urodynamic studies to monitor

adequacy of capacity and continence in all our patients after closure. If the bladder is too small for augmentation, the patient will need bladder substitution surgery (Inouye et al., 2014). Postoperatively, all patients after bladder surgery is immobilized for 4–6 weeks to allow the osteotomies to heal (Morales et al., 2016; Weiss et al, 2020; Inouye et al., 2014)

Successful primary bladder closure is the most important predictor of bladder growth and continence (Musleh et al., 2022; Baradan et al., 2011; Novak et al., 2010). Rates of continence after primary closure of bladder exstrophy ranges from 80 to 100%; but, the chance of developing an adequate bladder capacity decreases to 60% with failure of primary closure (Cervellione et al., 2015) Urinary continence is achieved either by an orthotopic urethra or an abdominal stoma constructed from ileum. Bladder augmentation is usually delayed until the patient is old enough to be taught methods of evacuation. There is no strict timing on when do we operate in these patients. It is often a patient centered decision. Some of our patients are lost to follow up. Admittedly, we would have wanted to know long term continence in our operated patients. The family should be prepared to participate as well, to help the child achieve continence and provide psychological support (Morales et al., 2016; Maruf et al., 2018). The median age for urinary continence in children is 11 years (Maruf et al., 2018). In some children that have continence problems even after exhausting surgical options, they are able to achieve continence using intermittent catheterization via a continent stoma or the urethra. Our choice for a stoma is Mitrofanoff appendicovesicostomy. In our institution, our patients are managed together with pediatric nephrology for their neurogenic bladder. We provide a multi-specialty approach not only to help the patient but provide support to families as well.

Neurologic issues caused by spinal cord involvement is the factor that is most likely to affect long term disability (Morales et al., 2016; Dutta, 2014) Early evaluation and management of these issues, along with close follow up are essential. Even though, this patient did not present with spinal cord anomalies both in the physical exam and in diagnostic evaluation we still keep the prognosis for fecal continence guarded until we are able to correct the anorectal malformation. In some cases we see, some patients achieve urinary continence but have fecal incontinence. The same is true the other way around.

Patients within the CE spectrum have been reported to have impaired overall quality of life. As discussed earlier, risk factors for impaired quality of life include urinary incontinence, genital dysfunction, dissatisfaction with genital appearance, and higher patient age. However, the existing

studies are insufficient to draw conclusions (Dellenmark-Blom et al., 2019). Historically, when CE was first recognized, affected infants often died of malnutrition and sepsis, even when successful surgical repair had been established. Presently, however, the mortality rate of this condition is less than ten percent (Novak et al., 2010; Dellenmark-Blom et al., 2019) In this case, the patient underwent successful surgical repair, and despite having neonatal pneumonia on admission, was discharged well. While our experience is limited compared to other higher volume centers that is purely dedicated to pediatric patients, we offer our approach to treating this complex anomaly.

Future short-term plan is to do urodynamic studies for bladder studies to assess bladder pressures and continence problems as the child grows. We plan to do multiple stage procedures; the second stage is do posterior sagittal anorectoplasty and use the end colostomy as the anal opening to treat the imperforate anus and genital reconstruction after genetic karyotyping and gender counseling. If phallic size does not increase in a few years, gender reassignment to a female will be probably appropriate. In the event that there will a grossly adequate penis size for correction, penile lengthening and reconstruction will be planned depending on the initial appearance of the phallus.

CONCLUSION

This case report describes a case of cloacal exstrophy who underwent successful staged surgical reconstruction and perioperative management. Although CE has been widely reported in literature, there are variety of approaches in the surgical management. Improvements in neonatal care and surgical techniques have improved survival and quality of life in these patients. Success rates have been reported on whether to do staged/delayed or primary repairs among authors. We still recommend careful patient selection for proposed procedures. Primary correction can be offered to term, healthy babies with manageable comorbidities. It is feasible to correct CE primarily with small bladder plates incorporating part of the bowel. Our proposed technique can be reproducible but it needs validation by other centers. Long term continence success rates is yet to be established in this type of procedure.

References

Admire, A. A., Greenfeld, J. I., Cosentino, C. M., Ghory, M. J., & Samimi, K. J. (2003). Repair of cloacal exstrophy, omphalocele, and gastroschisis using porcine small-intestinal submucosa or cadaveric skin homograft. *Plastic and Reconstructive Surgery*, 112(4), 1059-1062. https://doi.org/10.1097/01.PRS.0000076190.25804.B2

Baradaran, N., Cervellione, R. M., Orosco, R., Trock, B. J., Mathews, R. I., & Gearhart, J. P. (2011). Effect of failed initial closure on bladder growth in children with bladder exstrophy. *The Journal of Urology*, 186(4), 1450-1454. https://doi.org/10.1016/j.juro.2011.05.067

Cervellione, R. M., Mantovani, A., Gearhart, J., Bogaert, G., Gobet, R., Caione, P., & Dickson, A. P. (2015). Prospective study on the incidence of bladder/cloacal exstrophy and epispadias in Europe. *Journal of Pediatric Urology*, 11(6), 337-e1. https://doi.org/10.1016/j.jpurol.2015.03.023

Dellenmark-Blom, M., Sjöström, S., Abrahamsson, K., & Holmdahl, G. (2019). Health-related quality of life among children, adolescents, and adults with bladder exstrophy–epispadias complex: a systematic review of the literature and recommendations for future research. Q*uality of Life Research*, 28, 1389-1412. https://doi.org/10.1007/s11136-019-02119-7

Dutta, H. K. (2014). Cloacal exstrophy: A single center experience. *Journal of Pediatric Urology*, 10(2), 329-335. https://doi.org/10.1016/j.jpurol.2013.09.024

Gearhart, J., Di Carlo, H. N., Partin, A., Dmochowski, R., Kavoussi, L., & Peters, C. (2020). Exstrophy-epispadias complex: Campbell-Walsh-Wein urology. Philadel phia: Elsevier..

Inouye, B. M., Tourchi, A., Di Carlo, H. N., Young, E. E., & Gearhart, J. P. (2014). Modern management of the exstrophy-epispadias complex. *Surgery Research and Practice*, 2014. https://doi.org/10.1155/2014/587064

Jayman, J., Tourchi, A., Feng, Z., Trock, B. J., Maruf, M., Benz, K., ... & Gearhart, J. (2019). Predictors of a successful primary bladder closure in cloacal exstrophy: a multivariable analysis. *Journal of Pediatric Surgery*, 54(3), 491-494. https://doi.org/10.1016/j.jpedsurg.2018.06.030

Magahin, K., & Alviar, M. J. (2010). Rehabilitation of urinary incontinence following delayed bladder exstrophy repair in a female child. *Acta Medica Philippina*, *44(3)*. https://doi.org/10.47895/amp.v44i3.7762

Maruf, M., Kasprenski, M., Jayman, J., Goldstein, S. D., Benz, K., Baumgartner, T., & Gearhart, J. P. (2018). Achieving urinary continence in cloacal exstrophy: the surgical cost. *Journal of Pediatric Surgery*, *53(10)*, 1937-1941. https://doi.org/10.1016/j.jpedsurg.2018.02.055

Mathews, R., Wills, M., Perlman, E., & Gearhart, J. P. (1999). Neural innervation of the newborn exstrophic bladder: an immunohistochemical study. *The Journal of Urology*, *162(2)*, 506-508. https://doi.org/10.1016/S0022-5347(05)68616-8

Morales, G. M., Bustamante K.F., Rojas, I.M. (2016). OEIS Complex, A case Report. Journal of Intensive and Critical Care, *2(1).* https://doi.org/10.21767/2471-8505.100013

Musleh, L., Privitera, L., Paraboschi, I., Polymeropoulos, A., Mushtaq, I., & Giuliani, S. (2022). Long-term active problems in patients with cloacal exstrophy: a systematic review. *Journal of Pediatric Surgery*, *57(3)*, 339-347. https://doi.org/10.1016/j.jpedsurg.2021.08.020

Novak, T. E., Costello, J. P., Orosco, R., Sponseller, P. D., Mack, E., & Gearhart, J. P. (2010). Failed exstrophy closure: Management and outcome. *Journal of Pediatric Urology*, *6(4),* 381-384. https://doi.org/10.1016/j.jpurol.2009.10.009

Sadler, T. W. (2019). *Langman's Medical Embryology*. Lippincott Williams & Wilkins.

Sholklapper, T. N., Crigger, C., Haney, N., Khandge, P., Wu, W., Sponseller, P. D., & Gearhart, J. P. (2022). Orthopedic complications after osteotomy in patients with classic bladder exstrophy and cloacal exstrophy: A comparative study. *Journal of Pediatric Urology*, 18(5), 586-e1. https://doi.org/10.1016/j.jpurol.2022.09.005

Weiss D, Canning D, Borer J, Kryger J. (2020). *Bladder and Cloacal Exstrophy*. In: Holcomb G, Murphy J, St Peter S, ed. by. Holcomb and Ashcraft's Pediatric Surgery. Elsevier, Inc; 911-913.