

Bladder exstrophy-epispadias complex: patient and family psychosocial support

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Individuals with bladder exstrophy-epispadias complex (BEEC) and their families have unique needs best served by a multidisciplinary team of medical, surgical, and psychosocial providers. Global health initiatives to improve the overall health outcomes of this population are vital to share knowledge, cross-train providers, enhance collaboration partnerships, and address this population's unique needs. The Association for the Bladder Exstrophy Community (A-BE-C) and its volunteers have developed an initiative in Uganda to create a sustainable model of care in East Africa. Our dedicated group of volunteers and experienced providers intend to address the medical and psychosocial needs of the BEEC population. Given the complexity and psychosocial implications of these conditions, we firmly believe that the overall health outcomes of these patients rely on appropriate and life-long psychosocial services.

Therefore, our volunteer team consists of psychosocial providers and patient advocates. We have partnered with local physicians, nurses, social workers, community health workers, and disability rights advocates to create an annual comprehensive programme. In turn, our treatment plans are patient-centred, and the team collaborates to address medical and psychosocial needs simultaneously. In conjunction with the medical staff of nurses and physicians, the psychosocial team implements appropriate interventions to address barriers to care and self-image, improving independence and strengthening the family's hope and motivation. Our goals are to address and improve overall health outcomes, reduce shame and stigma, and support individuals with BEEC to reach their full potential.

Keywords: bladder exstrophy-epispadias complex, multidisciplinary care, psychosocial interventions, patient-centred care, global health

Introduction

The bladder exstrophy-epispadias complex (BEEC) are complex diseases that have life-long medical and psychosocial implications, including urinary incontinence, recurrent urinary tract infections, atypical genitalia, multiple surgeries throughout the lifespan, and kidney health concerns. Cloacal exstrophy (CE) is the most severe condition in this complex. It presents the individual with additional spinal, renal, reproductive system, and gastroenterological system abnormalities leading to faecal incontinence and ambulation challenges. Additional psychosocial implications include medical trauma from recurrent hospitalisations, surgeries and assessment, compromised self-image due to scars and abnormalities, mental health risks, interference in educational and career pursuits, and challenges related to marriageability. For individuals with BEEC, as with other chronic illnesses, the literature shows that patients affected are at risk for anxiety, depression, suicidality, isolation, and avoidance of intimacy.¹⁻⁵

Given the complexity of BEEC, a multidisciplinary approach is paramount. The A-BE-C is dedicated to servicing individuals with BEEC worldwide and is the only non-profit dedicated to this population.⁶ As part of their mission to reduce global health inequities, a 5–8 year initiative was developed in Uganda with Ugandan-based providers, stakeholders, and A-BE-C volunteers. The United States (USA)-based team – all of whom have substantial experience and expertise caring for this unique population – was created to include representation of urologic surgeons, orthopaedic surgeons, nursing, anaesthesiology, social work/behavioural health, and patient advocates. This initiative was developed to create a self-sustaining multidisciplinary programme for BEEC in Uganda.

Psychosocial principles

The main objectives of this mission's social work/behavioural health arm are to improve overall health outcomes, reduce shame and stigma, enhance community support, and for all individuals with BEEC "to realise their greatest life purpose and potential."⁶ This aligns with the global mission of social work as perfectly elaborated by the International Federation of Social Workers:

"Social work is a practice-based profession and an academic discipline that promotes social change and development, social cohesion, and the empowerment and liberation of people. Principles of social justice, human rights, collective responsibility and respect for diversities are central to social work. Underpinned by theories of social work, social sciences, humanities and indigenous knowledges, social work engages people and structures to address life challenges and enhance well-being."⁷

Care needs to be patient- and family-centred for individuals with BEEC to reach their full potential. Respecting their culture and community while fiercely advocating for their rights and safety is vital. Our initiative humbly aims to follow these principles in this collaboration. As with any international medical mission, deliberate communication, knowledge sharing, and determined partnership are fundamental in appropriately treating and caring for individuals in a cross-cultural context.

Psychosocial needs of the BEEC population

The diagnoses within the BEEC include highly intimate and sensitive aspects of hygiene and health. These rare conditions

can lead to urinary incontinence, faecal incontinence (CE), genital differences, and difficulties with sexual health/function. These issues can complicate the development of relationships, self-image, and personal development.¹⁻⁵ As such, the experience can be quite isolative and anxiety-inducing for both the affected person and their caregivers and families.⁸⁻¹³ Research on parents of children with BEEC revealed they experience significant levels of stress.¹⁴ Our team observed this anecdotally with our patients and the families cared for within this programme.

In the East African context, we have learned from our colleagues and patient families that these newborn babies with BEEC are often abandoned, neglected, seen as a curse or bad omen, and if not abandoned, isolated and hidden from their communities for years. We understand this is not unique to BEEC, but rather, it is a consequence of cultural ideas and norms regarding disability and anatomical differences worldwide.

Psychological trauma is the lasting emotional response to severe and distressing experiences. Traumatic events are experienced by an individual as physically or emotionally harmful, or life-threatening, and it has lasting adverse effects on the individual's functional, mental, physical, social, emotional, or spiritual well-being.¹⁵ For the caregivers and parents of those with BEEC, the presence of this difference in a newborn can be traumatising and invoke shock, grief, despair, confusion, and stress in parents and families. This can disrupt critical parent-child bonding.

For the parents, this trauma and additional stress (adjustment to diagnosis, financial demands, hospital visits, etc.) may lead to depression, suicidality, and rejection from their community. The parents' experience is transferred to the affected individual with BEEC, which can lead to years of psychological distress and trauma. With proper intervention, support, and education from the medical community, these families can adjust to a place of acceptance and advocacy. Connection to psychosocial partners in the community may allow the family to manage their trauma and grief in a transformative way.

Psychosocial interventions

This initiative relies on the partnership of Ugandan-based social workers, the A-BE-C, and a USA-based social worker (E Haddad) leading to peer-to-peer education. This reciprocal relationship allows Ugandan-based social workers to learn about this rare condition, including the medical implications of BEEC and the surgical treatments available to those with bladder exstrophy (BE). Meanwhile, the USA-based social worker learns about the nuanced

cultural and societal norms and community responses to those with disabilities. Furthermore, this facilitates the development of a sustainable and culturally appropriate approach to psychosocial support that acknowledges the challenges these families face in caring for their child with BEEC. Table 1 summarises the social work interventions provided during the mission/BEEC workshop.

Patient education day

This programme was created to address all aspects of care needed for the child with BEEC. We prioritise the integrity of the individual and family by promoting accessible educational material via interpreters regarding the unknown cause of BEEC, its surgical interventions, the condition's life-long implications, and recommendations for coping and advocating for their child and family. We share diagrams of the urological system, visuals of after surgery (primary closures, urinary diversions, etc.), and use a teaching doll with external fixators, stents, and catheters (Figure 1) for families to become acquainted with expectations after their child's bladder closure.

The A-BE-C utilises a puppet for medical play and as a teaching tool. Bunji, the puppet, was created specifically for this use, and it is complete with accurate urological and gastrointestinal systems. The teaching includes that the puppet, Bunji, was born with CE and underwent surgery for a Mitrofanoff and antegrade colonic enema (ACE) to obtain continence. As seen in Figure 2, this puppet has internal abdominal organs that can be used to describe enterocystoplasty and urinary stomas. It teaches children and caregivers about continence procedure options and post-surgery treatment plans/regimens (ACE flushes, CIC, bladder irrigation,



Figure 1: BE teaching doll; this doll has a nasogastric tube, right neck central venous catheter, right arm peripheral intravenous (IV), external fixator, suprapubic tube, and skin traction

Table 1: Summary of psychosocial interventions used in the BE population

Psychosocial intervention	Description
Patient education day	Medical and psychological education sessions with teaching tools to prepare families for treatment planning.
Standardised psychosocial assessments	Completed during the clinic and shared with the medical team to inform treatment planning.
Counselling and therapeutic support	Ongoing individual and group therapy throughout the workshop (medical decision-making support, pre- and post-surgery support/coping, clean intermittent catheterisation [CIC] teaching, etc.).
Case management	Mitigating barriers to medical care, supplies, education, career, etc.
School advocacy	Communication with school personnel to reduce discrimination and improve necessary accommodations.
Peer support	Formal and informal social support from other patients and families to improve outcomes and reduce isolation.



Figure 2: Bunji, the A-BE-C teaching puppet

etc.). Not shown in the images are continent catheterisable channel stoma, which is used to demonstrate the stoma's relative size and use.

The teaching tool in Figure 2 has representative internal anatomy consistent with CE. Though not shown in these images, the puppet has CE and both ACE and continent catheterisable urinary stomas, which help demonstrate the stoma size and catheterisation techniques to patients and their families.

The workshops in 2019 and 2023 have included local visual artist and disability activist Ritah Kivumbi and her fellow artists to provide therapeutic art activities for the families. Affected persons and volunteers are invited to share their stories, strength, and resilience with the patients and families. Rapport-building activities are facilitated to promote community engagement among the families.

Standardised psychosocial assessments

Psychosocial assessments are completed for each family in collaboration with the medical staff. These assessments inform family-centred treatment planning and collective decision-making regarding elective surgeries to ultimately attain urinary continence. Psychosocial readiness assessments are a universal quality standard for complex conditions. In this collaboration, surgeons and other medical providers rely on these vital psychosocial readiness assessments when making treatment recommendations and surgical plans.

Counselling and therapeutic support

Parents and caregivers of a newborn with BEEC may experience a great deal of isolation, anxiety, grief, and confusion. Early intervention is pivotal to ensure caregiver-child attachment. Counselling is provided to families at the workshop in conjunction with local behavioural health partners and our nursing colleagues. The fundamental pillars of therapeutic support are incorporated into care for families affected by BEEC: honesty, direct communication, active listening, validation, compassion, and respect.⁷

We utilise motivational interviewing techniques to encourage families to use their strengths in response to their fears and challenges.¹⁶

These strengths may include spiritual or religious beliefs, career aspirations, humour, artistic expression, etc. Cognitive behavioural therapy (CBT) interventions are also used to assist the patient and family with BEEC, acknowledge their problematic beliefs, and adopt adaptive coping skills.¹⁷ Culturally informed interventions and communication are imperative, highlighting the importance of the reciprocal relationship between USA-based and local psychosocial partners. These interventions must acknowledge the stigma and shame associated with mental health diagnoses, needs, services, and symptoms faced by the local population.

Psychosocial interventions before and during a medical intervention, such as CIC teaching for a school-aged child, may include relaxation techniques (breathing exercises and imagery). Exposure therapy is also utilised to introduce children and families to CIC gradually, as well as the postoperative presentation of the child (doll with external fixators and catheters after initial closure), urodynamics, etc. These interventions can be facilitated by the psychosocial professional, caregiver, nursing provider, or any medical team member to address the fear and pre-emptive anxiety a child has when preparing for medical interventions. Depending on the child and family's medical trauma history, they may need more or less interventions integrated into their treatment plan.

Case management

Partnerships with Ugandan-based community health personnel, medical doctors, school-based counsellors, and social workers are needed to holistically address the patient and family's needs and sustain the therapeutic relationship throughout the child's life. Case management aims to reduce barriers to accessing care, such as transportation, financial independence, educational pursuits, employment, and safety. The A-BE-C works closely with our Ugandan programme director (David Mukisa, B. Pharm, MPS) and local social workers to mitigate barriers. As the A-BE-C's mission is to address BEEC-specific medical needs, we refer families to other organisations to address financial, legal, or other needs.

School advocacy

Letters to advocate for non-discriminatory accommodations in the school setting related to urinary incontinence, CIC, and the need for regular restroom facility access is another critical component of our advocacy model. It is a worldwide challenge for individuals with BEEC to access appropriate education without discrimination or penalisation for their toileting needs; the situation is no different for our patients in Uganda. Some countries have laws to address this issue, and some communities have fewer formal means of managing these situations. We prioritise counselling families cared for in our BEEC workshop on ways to advocate for their children in the educational setting. Our local social work partners have been vital in delivering letters from the medical team to schools and attending meetings to facilitate accommodations for privacy and access and to reduce stigmatisation.

Peer support

Social support is informational, emotional, and instrumental assistance provided by others.¹⁸ This type of support can be formal or informal and benefits the individual and the group. Peer support in chronic illness reduces isolation and stress, protects against declining health, creates a sense of community, enhances feelings of hope, and promotes interpersonal learning and growth. Having a peer who relates to a medical situation can enhance confidence, medical adherence, and self-acceptance.¹⁸⁻²⁰ This fosters psychosocial well-being by protecting individuals from a sense of difference or not belonging.²¹ Given the rarity of the BEEC condition, the majority of these relationships usually rely on social media and long-distance communication.

Consequently, the one-week-long A-BE-C workshop allows families from across Uganda to build rapport with one another and connect intimately and powerfully. We encourage the families to support one another and facilitate small informal discussions, as appropriate. We witness our patients and their families experiencing these benefits during the workshop. Given the stigma of BEEC, access to peer support is part of the medical treatment plan. Peer mentorship and support enhance the family's coping and resilience during challenging times such as surgery, hospitalisation, and treatment planning. In addition to encouragement and emotional support, the families motivate one another to become advocates for their social acceptance in society.

Conclusion

Patients with BEEC are at risk for several long-term medical challenges, including recurrent urinary tract infections, chronic kidney disease, and even kidney failure. Psychosocial challenges related to urinary (and faecal) incontinence, genital differences, and body image challenges may lead to anxiety, depression, isolation, and the avoidance of intimate relationships, thus exacerbating the hesitancy to access healthcare. The consequences of these challenges are not only health-related but include financial hardship and a lack of access to education and meaningful employment. The most devastating risk for individuals with BEEC is abandonment upon birth and rejection from their communities. Despite these

obstacles, affected individuals can live a successful and healthy life with adequate interventions and support. Education starting with the caregivers to dispel misconceptions, provide peer support and community engagement, and offer therapeutic interventions can shelter individuals from these potential risks.

The A-BE-C has designed a comprehensive BEEC workshop in Uganda to help alleviate the risks associated with BEEC. We provide multidisciplinary interventions that we expect will lead to positive outcomes. The project we have created in collaboration with local partners is meant to be sustainable in Uganda and prioritises the integrity and well-being of the children and adults with BEEC and their families.

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Dedication

This paper is dedicated to our late colleague and friend, Martin Situma, MD.

Conflict of interest

The authors declare no conflict of interest.

Ethical approval

The authors declare that this submission follows the Responsible Research Publication Position Statements principles developed at the Second World Conference on Research Integrity in Singapore in 2010. This work is exempt from Institutional Review Board approval because it did not directly involve human subjects.

References

1. Anderson DL, Murray CD, Hurrell R. Experiences of intimacy among people with bladder exstrophy. *Qual Health Res.* 2013;23:1600-12. <https://doi.org/10.1177/1049732313509409>.
2. Deans R, Liao L-M, Wood D, Woodhouse C, Creighton SM. Sexual function and health-related quality of life in women with classic bladder exstrophy. *BJU Int.* 2015;115(4):633-8. <https://doi.org/10.1111/bju.12811>.
3. Dellenmark-Blom M, Sjöström S, Abrahamsson K, Holmdahl G. 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. *Qual Life Res.* 2019;28(6):1389-412. <https://doi.org/10.1007/s11136-019-02119-7>.
4. Baird AD. Exstrophy in the adolescent and young adult population. *Semin Pediatr Surg.* 2011;20(2):109-12. <https://doi.org/10.1053/j.sempedsurg.2010.12.006>.
5. Pennison MC, Mednick L, Rosoklija I, et al. Health related quality of life in patients with bladder exstrophy: a call for targeted interventions. *J Urol.* 2014;191(5 Suppl):1553-7. <https://doi.org/10.1016/j.juro.2013.09.089>.
6. bladderexstrophy.com [Internet]. Association for the Bladder Exstrophy Community; 2022. Available from: www.bladderexstrophy.com. Accessed 14 April 2023.
7. ifsw.org [Internet]. International Federations of Social Workers; 2023. Available from: www.ifsw.org. Accessed 14 April 2023.
8. Bujons A, Lopategui DM, Rodriguez N, et al. Quality of life in female patients with bladder exstrophy-epispadias complex: long-term follow-up. *J Pediatr Urol.* 2016;12(4):210.e1-6. <https://doi.org/10.1016/j.jpuro.2016.05.005>.
9. Da Cruz JAS, de Mattos B, Srougi M, et al. Quality of life in young adult patients treated for bladder exstrophy. *Cent European J Urol.* 2016;69(2):221-4. <https://doi.org/10.5173/cej.2016.715>.
10. Di Grazia M, Pellizzoni S, Tonedatti LG, Rigamonti W. Qualitative questionnaire on the psychosocial wellbeing of mothers of children with BEEC. *J Pediatr Urol.* 2017;13(1):55.e1-6. <https://doi.org/10.1016/j.jpuro.2016.07.015>.

11. Reiner WG, Gearhart JP. Anxiety disorders in children with epispadias-exstrophy. *Urology*. 2006;68(1):172-4. <https://doi.org/10.1016/j.urology.2006.01.056>.
12. Holmdahl G, Dellenmark-Blom M, Nordenskjöld A, Sjöström S. Health-related quality of life in patients with the bladder exstrophy-epispadias complex and relationship to incontinence and sexual factors: a review of the recent literature. *Eur J Pediatr Surg*. 2020;30(3):251-60. <https://doi.org/10.1055/s-0040-1713178>.
13. Ebert AK, Schott G, Bals-Pratsch M, Seifert B, Rösch WH. Long-term follow-up of male patients after reconstruction of the bladder-exstrophy-epispadias complex: psychosocial status, continence, renal and genital function. *J Pediatr Urol*. 2010;6(1):6-10. <https://doi.org/10.1016/j.jpuro.2009.06.002>.
14. Mednick L, Gargollo P, Oliva M, Grant R, Borer J. Stress and coping of parents of young children diagnosed with bladder exstrophy. *J Urol*. 2009;181(3):1312-6; discussion 1317. <https://doi.org/10.1016/j.juro.2008.10.051>.
15. Substance Abuse and Mental Health Services Administration. SAMHSA's concept of trauma and guidance for a trauma-informed approach. Rockville: SAMHSA's Trauma and Justice Strategic Initiative; 2014.
16. Erickson SJ, Gerstle M, Feldstein SW. Brief interventions and motivational interviewing with children, adolescents, and their parents in pediatric health care settings: a review. *Arch Pediatr Adolesc Med*. 2005;159(12):1173-80. <https://doi.org/10.1001/archpedi.159.12.1173>.
17. Tok ESS. Cognitive behavioral therapy principles in children: treatment of internalizing disorders. In: Şenormancı Ö, Şenormancı G, editors. *Cognitive behavioral therapy and clinical applications*. IntechOpen; 2018. p. 1-25. <https://doi.org/10.5772/intechopen.71932>.
18. Finlay KA, Elander J. Reflecting the transition from pain management services to chronic pain support group attendance: an interpretative phenomenological analysis. *Br J Health Psychol*. 2016;21(3):660-76. <https://doi.org/10.1111/bjhp.12194>.
19. Scheel A, Beaton A, Okello E, et al. The impact of a peer support group for children with rheumatic heart disease in Uganda. *Patient Educ Couns*. 2018;101(1):119-23. <https://doi.org/10.1016/j.pec.2017.07.006>.
20. Scheurer D, Choudhry N, Swanton KA, Matlin O, Shrank W. Association between different types of social support and medication adherence. *Am J Manag Care*. 2012;18(12):e461-e7.
21. Kaufman N. Peer-to-peer support helps people improve their chronic condition therapy outcomes [Internet]. Canary Health Chronic Disease Programs; 2022. Available from: <https://www.canaryhealth.com/peer-to-peer-support-improves-chronic-condition-therapy-neal-author/>. Accessed 9 January 2023.