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CASE REPORT
Craniofacial/Pediatric

Effective Collaboration in the Surgical Management of Macroglossia in Beckwith–Wiedemann Syndrome

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Summary: Beckwith–Wiedemann syndrome (BWS) is a complex congenital overgrowth disorder necessitating a multidisciplinary approach for effective management. A 5-year-old Saudi girl with BWS received comprehensive care involving various specialists, including a plastic surgeon who performed a keyhole technique tongue reduction to address macroglossia. The intervention resulted in significant improvements in speech and quality of life, with no postoperative complications. Intensive speech therapy further enhanced speech development. This case report emphasizes the importance of a multidisciplinary approach and the critical role of the plastic surgeon in managing BWS patients with macroglossia to achieve optimal outcomes. *(Plast Reconstr Surg Glob Open 2024; 12:e5635; doi: 10.1097/GOX.0000000000005635; Published online 8 March 2024.)*

Beckwith–Wiedemann syndrome (BWS) is a rare genetic overgrowth disorder, affecting approximately one in 10,000 to one in 13,700 live births. This syndrome is characterized by various clinical features, including macroglossia (enlarged tongue), abdominal wall defects, overgrowth, and an elevated risk of embryonal tumors. Macroglossia, present in about 90% of BWS cases, can lead to functional, aesthetic, and psychological challenges due to skeletal muscle hyperplasia.[1–4]

Managing BWS is intricate, necessitating a multidisciplinary approach involving pediatricians, endocrinologists, geneticists, nephrologists, orthodontists, pediatric dentists, and plastic surgeons. This case report focuses on the collaborative management of a 5-year-old girl with BWS, emphasizing the challenges associated with macroglossia and its impact on speech, oral health, and overall quality of life.

The case underscores the significance of a coordinated, multidisciplinary approach in addressing the complex needs of BWS patients, with a particular emphasis on the crucial role of plastic surgeons in enhancing patients' quality of life.

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CASE PRESENTATION

A 5-year-old Saudi girl with BWS was referred to our team for management. Diagnosed at 1 year due to macroglossia, her medical history includes BWS, polycystic kidney disease, and a language disorder. Despite lacking regular dental visits, she displayed poor oral hygiene and Frankel I behavior during treatment. Clinically, she presented macroglossia, spaced dentition, carious teeth, and an anterior open bite. Caries Risk Assessment categorized her with a high caries risk.

MANAGEMENT

A comprehensive care plan for our 5-year-old patient with BWS was executed by a multidisciplinary team, including an endocrinologist, pediatrician, geneticist, pediatric dentist, orthodontist, nephrologist, plastic surgeon, and speech pathologist. Initially referred at age 5, the patient underwent simultaneous dental rehabilitation and tongue reduction surgery using the keyhole technique, preserving lateral neurovascular bundles.[5–8]

Under general anesthesia with nasal intubation, the surgical procedure began. Strategic markings on the dorsal tongue outlined the resection area, and using the keyhole technique, the tongue's length and width were reduced. Local anesthesia (1% lidocaine with adrenaline) was administered, and an incision through the superior longitudinal, vertical, and transverse muscle layers was made, removing a small portion of the inferior longitudinal muscle.

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After lateral flap alignment, the revised tongue size was examined, ensuring passive oral cavity closure without obstructions. Additional resection targeted intrinsic muscles for optimal size reduction. After resection and hemostasis, muscle layers were sutured (3/0 Vicryl), and mucosa was horizontally closed (4/0 Vicryl rapid). Approximately 30% of the anterior two-thirds of the tongue volume was selectively removed, addressing skeletal muscle hyperplasia and placing postoperative tongue size within the normal pediatric range.

Orthodontic management addressed the enlarged tongue, anterior open bite, incompetent lips, and incisor bowing. Regular follow-up with the orthodontist monitored occlusal growth, considering dynamic changes induced by tongue volume reduction.

Postsurgery, significant speech improvements were noted by the speech-language pathologist, with enhanced articulation, phonation, and speech intelligibility (measured using standardized speech assessment scales[9]). A structured interview confirmed the patient's ability to discern all four primary tastes, indicating preserved taste sensation.

Structured follow-up appointments were scheduled immediately postoperation and at regular intervals. These sessions assessed oral health, speech progression, and overall adaptation to changes. The patient remains consistently under active follow-up for long-term outcome monitoring and adjustments to the care plan. Refer to Supplemental Digital Content 1 for additional details on the intervention timeline. (**See table, Supplemental Digital Content 1,** which shows timeline of multidisciplinary management in a patient with Beckwith–Wiedemann syndrome. **http://links.lww.com/PRSGO/D82**.)

FOLLOW-UP

After surgery, rigorous postoperative assessments were conducted at specific intervals to assess recovery and intervention efficacy. Immediate postoperative review revealed no complications, intact dental restorations, and improved oral hygiene, likely due to reinforced dental care education. Gustatory function preservation was confirmed, with the patient exhibiting normal taste sensation for all primary tastes. Positive behavioral changes were noted during subsequent dental visits, indicating increased comfort with treatments.

At 1 month postoperation, monitored by a speech-language pathologist, weekly speech therapy sessions and daily home exercises led to significant improvements in articulation, phonation, and speech intelligibility (measured through standardized scales[9]).

By the 6-month follow-up, the patient maintained good oral hygiene, intact dental restorations, and normal taste sensations. Enrolled in a long-term follow-up program, sessions included oral hygiene reinforcement, topical fluoride application, dietary evaluations, dental restoration checks, and orthodontic assessments.

Future orthodontic plans involve reevaluation around age 7–8, determining interventions based on growth. Figures 1–4 illustrate the patient's remarkable progress during long-term follow-up, showcasing the positive impact of our multidisciplinary intervention.

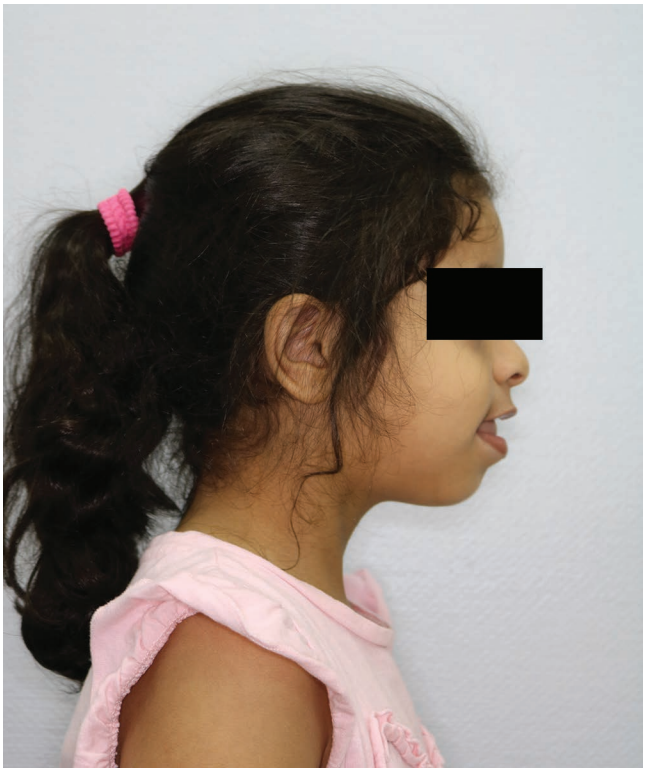

Fig. 1. The patient before the management of her condition.

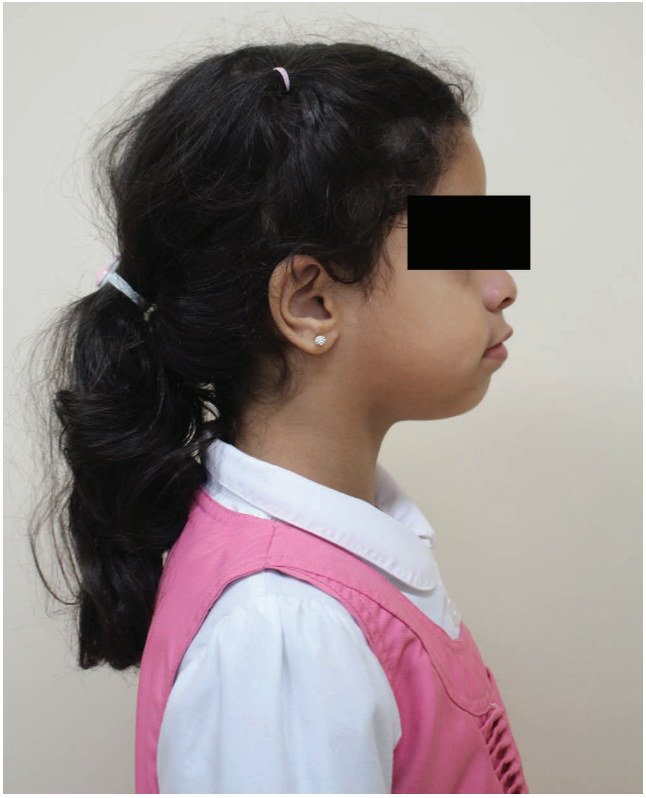

Fig. 2. The patient during her routine follow-up after the management of her condition.

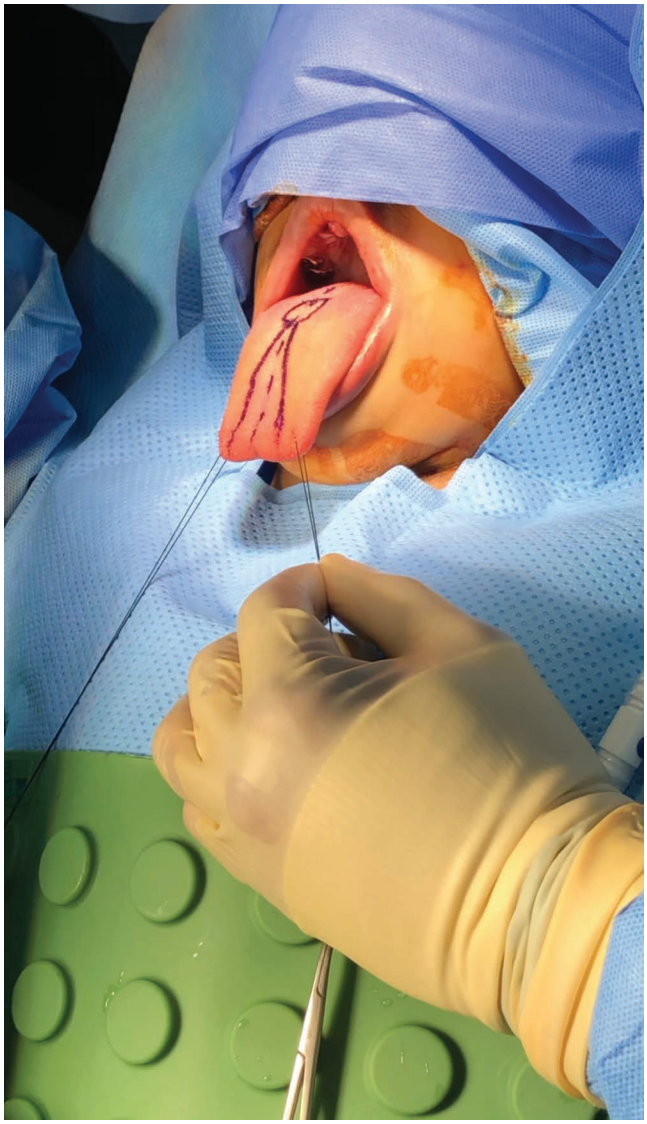

Fig. 3. Intraoperative markings of the keyhole technique.

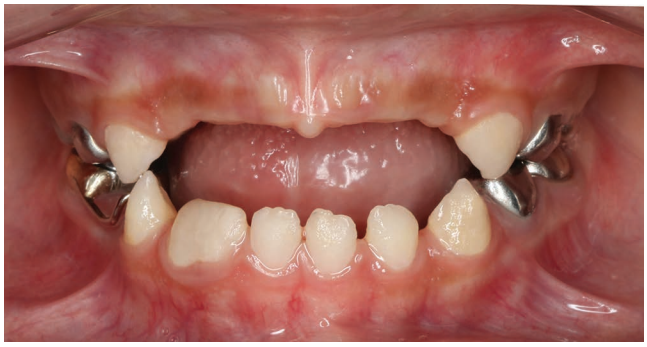

Fig. 4. Intraoral improvement of the patient during routine follow-up.

DISCUSSION

The complex needs of individuals with BWS demand a holistic, interdisciplinary management approach, encompassing dental and orthodontic care, speech therapy, and surgical interventions.[3,5] In this context, the plastic surgeon plays a pivotal role not only in executing surgical procedures but also in the comprehensive management of the patient's condition, as exemplified in our case report.[5–8,10]

Macroglossia, a characteristic feature of BWS, often leads to functional impairments in speech, swallowing, and chewing, with potential psychological impact due to social stigmatization.[7] In our presented case, concerns raised by the patient's mother, including a large tongue, drooling, and speech difficulties, underscored the need for surgical intervention to enhance the child's condition and facilitate more effective speech therapy.[5,7]

Led by the plastic surgeon, the surgical approach used the keyhole technique, combining anterior wedge resection, and central tongue reduction. Preserving the lateral neurovascular bundles, this successful procedure significantly improved speech capabilities and overall quality of life.[6–8,10]

The multifaceted roles of plastic surgeons extend beyond surgery, encompassing collaboration with other specialists in the multidisciplinary team to develop personalized treatment plans optimizing function, aesthetics, and overall well-being.[4] Active involvement in postoperative care ensures smooth recovery and successful integration of surgical interventions within the broader management plan.[5]

Lessons from this case stress the importance of early diagnosis and intervention in BWS, emphasizing the significance of a multidisciplinary team involving various specialists. Regular follow-ups are crucial for tracking growth and development, and assessing ongoing interventions.[2,4]

Educating parents about the syndrome, potential complications, and the importance of adherence to follow-up visits and treatment plans is imperative for optimal care and support for the child.[4,7] Furthermore, promoting oral health through good hygiene, dietary counseling, and regular dental visits is crucial in preventing complications and enhancing overall quality of life.[5,6]

CONCLUSIONS

The plastic surgeon plays a pivotal role in BWS management, particularly in addressing macroglossia, a key aspect of the comprehensive treatment framework. Their expertise in refined surgical techniques and active involvement in the multidisciplinary team significantly enhance functional abilities, aesthetics, and overall quality of life.

Beyond the operating room, the plastic surgeon's contribution extends to holistic postoperative care, consistently monitoring and addressing concerns or complications. As our understanding of BWS grows and management evolves, the collaboration of diverse specialists, with plastic surgeons at the forefront, remains essential. This team-based approach ensures comprehensive, patient-centered care, optimizing functional, aesthetic, and psychosocial outcomes for individuals with BWS.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

PATIENT CONSENT

Written patient and parents' consents were taken for scientific publication.

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This case report confirms to the Declaration of Helsinki.

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