The Versatility of Tissue Expansion in Providing Primary Wound Coverage in Conjoined Twins Separation

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ABSTRACT

The successful separation of conjoined twins entails providing enough soft tissue for reconstruction and coverage of the resulting defects. A variety of techniques have been employed for this objective, but most have its peculiar drawbacks. Herein, we report successful primary skin closure at the time of separation of two sets of thoraco-omphalopagus and pyopagus twins utilizing single tissue expanders without complications. We also report a unique method of using pedicled flaps harvested from the pseudocapsule surrounding the implant to act as a second layer reinforcing the closure over the underlying vital structures. Even though tissue expansion is a two-stage procedure, it carries a lot of benefits. It provides the anesthetic team with a golden opportunity to rehearse prior to the actual separation and allows the pediatric surgeons to perform their abdominal laparoscopy and any other necessary investigations under anesthesia. Furthermore, having ready-to-use expanded flaps decreases the overall operative time during the critical stage of separation.

INTRODUCTION

Conjoined twins represent a unique congenital anomaly that occurs in approximately 1 in 50,000 births [1]. However, as up to 60% are stillborn, the true incidence of live-born conjoined twins approaches 1 in 200,000 live births [2]. In Africa the incidence has been quoted as high as 1 in 14000 deliveries, thus suggesting an increased frequency in blacks [3]. Females predominate with a 3:1 ratio. Conjoined twins are classified according to the most predominant site of connection, with thoracopagus and omphalopagus (thoraco–omphalopagus) accounting for approximately 74% of cases, with fusion from the anterior thorax to the umbilicus. The union can be in the frontal, transverse, or sagittal plane. A common pericardial sac is present in 90% of thoracopagi and conjoined hearts are seen in 75%. The term "pyopagus" is given to twins joined at the sacrum and accounts for approximately 18% of cases, the sacra are fused and the twins may share a portion of the spinal cord. In addition, the rectum and perineal structures are usually fused. Ischiopagus twins (i.e., joined at the ischium) represents 6% while Craniopagus twins (i.e., joined at the head) occurs in 2% of conjoined twins. The term 'parapagus' has been introduced to indicate extensive side-to-side fusion [2,3].

Treating conjoined twins is often extremely complex and experience with large numbers has been traditionally restricted to a few centers worldwide. Nevertheless, surgical experience with conjoined twins has been reported recently in many parts of the world, such as China, Saudi Arabia, and New Zealand [4-6]. In conjoined twins with non life-threatening joined structures, there is generally good prognosis, with time for investigations and planning of the operative separation [7]. Improved recent survival is probably the result of the availability of more accurate imaging studies and better anesthetic and operative techniques, with great emphasis on performing immediate reconstruction whenever possible [8]. If the shared anatomy allows for separation and viability of both twins as in the case of pyopagus and thoraco-omphalopagus twins, the problem of primary wound closure emerges. This is especially true, if the area of attachment is large. More than a technical problem, difficulties in providing skin coverage can threaten the lives of those infants [9].

The ultimate skin deficiency is determined by the size of the fusion area. In a successful separation, the deficiency equals the cross-sectional area of fusion multiplied by two, because coverage must be provided for both infants. A variety of techniques have been used to provide skin, but most have its peculiar drawbacks. Skin flaps can be extensively elevated from the abdominal wall and advanced for primary closure. The flaps are basically random in nature and are almost always sutured under great
Tissue expansion has been introduced as a favorable method, but its successful application to the conjoined twins is seldom reported [9,13,15-17]. Filler [16] has described the technique of utilizing skin expanders within the peritoneal cavity in order to enlarge the abdominal wall in the separation of ischiopagus twins, while O’Neil [8] used them in the subcutaneous location in three instances to subsequently achieve tension-free closure. Moreover, tissue expander use in children has been reported with high complication rates of up to 26%, with infections being the most common reason for expander removal [17].

The aim of this article is to present the challenges that our team have faced in providing primary closure for two sets of conjoined twins using the technique of tissue expansion.

**PATIENTS AND METHODS**

Two sets of female conjoined twins were managed at Ain-Shams University Hospitals and Ahmed Maher Teaching Hospital. The first set was a pyopagus twin while the second was a thoraco-omphalopagus twin.

**Clinical data:**

Full-term female pyopagus conjoined twins were born on December 20th of 2004 through normal vaginal delivery and referred to Ain-Shams University Hospitals. The infants were joined back to back. Initial evaluation confirmed their fusion at the sacral level with abnormal perineal anatomy as they shared one anus. However, each baby had a separate urethra (Fig. 1). Both infants had normal lower limbs neurologically with normal movements and no wasting. Other gross abnormalities were ruled out at the time of initial examination. Imaging studies of the twins were undertaken to further assess the shared structures. A retrograde contrast study identified that the twins’ intestine was connected to the single anus; therefore a colostomy was planned for at the time of separation. The plain X-ray demonstrated a "U"-shaped fusion of two vertebral columns at level of S3. Appropriate cross-sectional imaging, including CT & MRI were performed to further delineate the pyopagus anatomy. This revealed a "U"-shaped fusion of two lower spinal cords. The thecal sacs were in continuity, but there was no evidence of fusion of neural tissues. Evaluation of the genitourinary tract revealed four normal kidneys, ureters and separate bladders.

A multidisciplinary management team was put together and comprised neonatologists, pediatric surgeons, neurosurgeons, plastic & reconstructive surgeons, anesthetists and nursing. A detailed first multidisciplinary planning meeting was held, with representation from all specialties involved in the care of these children. Team meetings followed on regular basis to discuss their progress, diagnostic, and therapeutic requirements. Since the joined structures were not life threatening with an expected good prognosis after separation, it was decided that the infants were allowed to be fed normally and to gain weight for six months prior to any attempt of surgical intervention, allowing for growth and more physiological stabilization. As the fused area at the lower back was large measuring 8.5cm vertically and 9.0cm transversely (Fig. 2), it was decided as a preliminary to insert a tissue expander beneath the joining skin bridge in order to facilitate skin closure at the time of separation.

**Surgical technique:**

A single soft-based tissue expander (Nagor”, Nagor Limited, British Isles), 200 cc, rectangular in shape, with dimensions of 8.5 x 5 x 5cm, was placed subcutaneously at the furrow of fusion site...
with its longitudinal axis being parallel to the longitudinal axis of fusion. The plan was to allow simple skin advancement for one twin and an expanded transverse back flap for the other. The incision to insert the expander was remote and radial. It was placed well away from anus and urethra to prevent their injury and to avoid being contaminated. An internal port was used to prevent the higher risk of infection if external ports were used and also to avoid pulling or damaging by the babies. The inflation dome was placed as remote as possible and was well palpable, so there was no chance of being covered by the balloons when over expanded (Fig. 3). Expansion was started once access incision has healed within a couple of weeks. It continued on regular basis as twice weekly with small increments of saline. Over expansion was performed and up to 500cc was inflated after ten weeks to gain extra tissues than calculated for (Fig. 4). The expander was left for an additional two weeks after full expansion to allow for maturation of its surrounding capsule in order to avoid retraction of the expanded skin at the time of separation and to enable dissecting a flap from the pseudocapsule to be used as reinforcing layer for dural closure.

The second-stage operation was performed over three months following the first stage. Pediatric surgeons, neurosurgeons and plastic surgeons undertook the definitive separation. The infants were placed on the operating table in the lateral position, back to back to enable removal of the tissue expander and marking out the appropriate skin flaps. A preliminary incision on the side of the balloon allowed its removal leaving a well-defined capsule with insertion of gauze to maintain the flaps expanded till separation has been completed. On deepening the incision the spinous processes and shared posterior sacral bony elements of each infant were exposed. The neurosurgical team performed the spinal cord separation including dural tube closure. The pediatric surgeons carried out the perineal separation, essentially separating the rectum of each twin and fashioning a separate vagina for each. Once finally separated one twin was taken back to the adjacent operating table and both were repositioned prone for completion of their operative repair. Part of the pseudocapsule tissue was incised and dissected as a pedicled flap and used to enhance the two dural closures [rather than using lumbar fascia or fascia lata grafts]. On completion of repair, there was no evidence of obvious cerebrospinal fluid (CSF) leak. Primary skin closure for the skin defects was possible in each infant with the expanded skin flaps (Fig. 5). They were discharged in a good condition only one week after the separation.

**The second pair (The Thoraco-omphalopagus twins):**

**Clinical data:**

Full-term female thoraco-omphalopagus twins were born in 28 January, 2006 by elective caesarean section at Ahmed Maher Teaching Hospital. They were joined from the mid sternum to the umbilicus (Fig. 6). The area of fusion measured 12.5cm vertically and 7cm transversely. Studies of the twins were undertaken to assess the shared structures. Plain X-rays, ultra sonogram, CT with contrast and MRI studies showed that the twins had separate bowels, hearts, lungs, intra-hepatic portal system, arterial system, venous system and normal kidneys. There were joined livers with crossover hepatic blood flow in-between. The abdominal ultrasonography revealed gall bladder in one infant and porta-hepatis in the second infant. Echo-Cardiography, revealed atrial septal defect (ASD) and patent ductus arteriosus (PDA) in the first infant and patent foramen ovale (PFO) and PDA in the second infant, with attached common pericardial sac and continuous muscular texture of diaphragm. A management team was assembled and the infants were fed normally for 6 months before any surgical intervention, allowing for growth and physiological stabilization. During this time the anticipated problems of skin coverage were considered.

**Surgical technique:**

While achieving tension free primary wound closure at the time of separation being our goal, tissue expansion was selected. Adopting the same surgical principles as in the first set of twin, the first stage surgery was performed at the age of 6 months. A single soft-based tissue expander (Nagor®, Nagor Limited, British Isles) measuring 8.5cm in length x 5cm in width x 5cm in height, with a total capacity of 200cc, was used. The pocket was dissected subcutaneously and was chosen to be in the joining bridge over the fused thoracic bony cage. This allowed the expander to rest on bone for maximal expansion yield. It also provided the expanded skin being adjacent to the site of expected defect at the thoracic and upper abdominal regions at the time of separation (Fig. 7). Furthermore, it allowed delivering the expander last during the separation stage. During this stage, the pediatric surgeons performed an abdominal laparoscopy from a separate access incision. The expansion process was started once access incision

has healed within a couple of weeks where small amounts of normal saline were injected twice weekly on regular basis. A good expansion volume was achieved by the time we reached a total of 550cc (Fig. 8). Two extra weeks were allowed to pass after full expansion and before the actual separation stage to allow for maturation and to avoid expanded skin retraction later on.

The actual stage of separation and definitive primary repair was performed after three months. The infants were placed in the lateral position with the balloons on the underside to gain easy access to the conjoined livers and the biliary system. Abdominal incision was carried out first along the site of fusion followed by hepatic separation after intraoperative mapping was done. The incision was carried further cephalic with delivery of the expander. The hearts were completely separate anatomically and functionally but were fused partially at the pericardium. This common pericardial sac was opened and repaired primarily. The remaining joining abdominal wall at the undersurface was incised completing the separation. One twin was taken to the adjacent operating table and both were repositioned supine for completion of their operative repair. A double-layered mesh (inner absorbable teflon and outer prolene) was used to reconstruct the defective rib cage at the site of fusion in both twins. Over which a pseudocapsule flap was dissected from the undersurface of the expanded skin flap and rotated to reinforce the reconstruction by a second layer closure (Fig. 9).

Adequate primary skin closure was possible in both infants (Fig. 10) by expanded skin flaps over the thoracic and superior abdomen. The remaining part of the abdominal wall defect was closed directly without tension. On postoperative day 10, the infants were discharged from the hospital.

RESULTS

The expansion was well tolerated by both infants. The two sets of conjoined twins were successfully separated with primary skin closure in spite of the large shared area of fusion. The skin approximation was accomplished under no tension with good cosmetic results. There was never any evidence of infection or ischemic compromise of the expanded skin flaps.

There was no CSF leak postoperatively in the pyopagus twins, denoting good dural closure by the enforcing flap harvested from the pseudocapsule formed around the tissue expander. In the thoraco-omphalopagus twins, the pseudocapsule flap provided a good reinforcing additional layer for thoracic wall closure.

The recognized problems encountered in traditional methods of wound closure at the separation (including wound breakdown, infection, ventilatory compromise and obstruction to venous return resulting from tight abdominal skin closure) were avoided through the use of this technique. All vital internal organs and structures (including the hearts and spinal cords) were well protected and covered by the expanded skin flaps.

All the wounds healed well and no further complications occurred at 1-year follow-up and the twins were progressing well under the care of the multidisciplinary team and physiotherapists.

DISCUSSION

Conjoined twinning is one of the most fascinating human malformations. Their management remains exciting and challenging. They have been classified according to the most important site of attachment into craniopagi, thoracopagi, omphalopagi, ischiopagi and pyopagi [18]. Thoracopagi are the commonest type accounting for about 40% of the cases followed by omphalopagi (34%). Pyopagus conjoined twins are rarer, accounting for about 10-18% of all conjoined twins which gives an incidence of about one in 1,000,000 live births of this particular type. This classification was addressed by Spencer [19] and although eight different types of conjoined twins have been described, each set has its unique features that necessitate thorough and detailed preoperative investigations. According to the work of Spencer [19], the conjoined twins are identified by the external examination of the area of union except in the case of thoracopagus and omphalopagus forms of twins, which may exhibit identical external musculoskeletal union from sternal notch to the umbilicus but have very different internal unions. The importance of differentiating between these two is their vastly different prognosis. Thoracopagus should be reserved for those with some degree of union of the hearts, usually very complex and almost always not separable [19,20]. Omphalopagus twins do not exhibit cardiac union, but almost always have liver union, sometimes fused sternal cartilage (in 57% of cases) [20,21] and on occasion some fusion of portions of the gastrointestinal tract, (e.g., duodenum or Meckels diverticulum). These twins are usually separable and will survive if other anomalies are not life limiting [21-23]. In our patients the findings in the second pair were consistent with the omphalopagus category.
Fig. (1): Pyopagus twin; (a) front view demonstrating their fusion back to back at the sacral level. (b) caudal view demonstrating abnormal perineal anatomy.

Fig. (2): The anticipated wound following separation was large as the fused area at the lower back measured 8.5 cm vertically and 9.0 cm transversely.

Fig. (3): The expander was placed beneath the joining skin valley parallel to the longitudinal axis of fusion through a radial incision. The inflation dome (red circle) was placed remotely over the caudal end of the rib cage of one of the twins.

Fig. (4): Over expansion was performed and up to 500 cc was inflated after 10 weeks (a) side view (b) top view.
Fig. (5): Primary skin closure was possible for both infants by simple skin advancement in twin (a) and an expanded transverse back flap for twin (b).

Fig. (6): Thoraco-omphalopagus twin joined from mid-sternum to umbilicus.

Fig. (7): The expander placement was designed to be subcutaneously over the fused sternum through a radial incision (red line). The inflation dome (red circle) was placed remotely over the rib cage of one of the twins.

Fig. (8): Comparing the area of fusion over the thoracic cage before (a) and after (b) overexpansion.
Pyopagus twins commonly share the gluteal region, terminal spine, gastrointestinal, urological and reproductive systems to variable degrees. Although the joined structures are not life threatening, the extent of attachment as well as the presence of other anomalies including spinal cord and vertebral anomalies are of paramount importance [26,27]. About 50% of pygopagus twins have anomalies unrelated to the classic fused organs including a high incidence of vertebral anomalies [26]. Our pyopagus pair shared one anus and a "U"-shaped fusion of the two vertebral columns at level of S3 with the thecal sacs in continuity, but there was no evidence of fusion of neural tissue.

The first successful separation of conjoined twins was performed in 1689 by Johannes Fabio [24]. In an extensive review, Hoyle in 1990 [25] analyzed 167 attempts at surgical separation of conjoined twins and quoted an overall survival rate of 64%. Thoracopagus, craniopagus and omphalopagus were associated with the highest mortality rates (51%, 48% and 32%, respectively), while lower mortality rates occurred with ischiopagus (19%) and pygopagus (23%) twins [25]. The outcome of separation of pyopagus and thoraco-omphalopagus twins in the modern era is very successful owing to the nature of the fusion. Herein, we report successful separation of two sets of

Fig. (9): Raising a pedicled pseudocapsule flap from the under surface of expanded skin (a). This flap was used as a second layer closure to reinforce the reconstruction over the synthetic mesh (b).

Fig. (10): Primary skin closure was achieved in both infants by expanded skin flaps over the thoracic and superior abdomen. The remaining part of abdominal defect was closed directly without tension.
thoraco-omphalopagus and pyopagus twins utilizing single tissue expanders for primary skin closure and a unique method using a flap harvested from the pseudocapsule surrounding the implant as a second layer reinforcing the closure over the underlying vital structures.

Timing of separation of conjoined twins is often debated. In situations were the physiological status of the infants is secured, delay of separation allowing the infants to grow and stabilize further is preferred. On the other hand, if separation is delayed much beyond 1 year, there is some evidence to suggest that the twins may have difficulty developing a separate identity [9,23]. McDowell et al. (2003) [28] estimated survival rates for pyopagus separation to be 50% when performed in the neonatal period compared with 90% if the operation was delayed at least 6 months. Recent separations described by Hockley et al., 2004 [7] and Feiggen et al., 2004 [29] support this outcome. Thus, if feasible, surgery should be delayed until physiological reserve can be maximized and the appropriate imaging and extensive operative planning formulated. In our patients, expander placement was elected to be around six months. This allowed time for proper feeding to reach an acceptable skin & subcutaneous soft tissue thickness that would withstand the tension created by the underlying expansion thus avoiding unnecessary implant exposure. The final separation was planned around the age of nine months coinciding with the most appropriate time for separation of such cases.

Frequently, the most challenging portion of successful separation of conjoined twins rests with the plastic surgery team and their capabilities in providing enough soft tissue for reconstruction and coverage [8,30,31]. This becomes of paramount importance when the area of attachment is large and in the presence of underlying vital organs [32]. A variety of techniques have been proposed to achieve primary skin closure including skin grafts, tissue flaps, and prosthetic patches. However, none was satisfactory and most of them had highlighted long-term problems with sinuses and chronic infections [10,32-37]. Therefore, we adopted a staged surgical approach as an alternative management strategy in our cases. In the first stage, insertion of tissue expanders subcutaneously at age of six months and definitive separation on an elective basis was performed after three months. This technique has been used sporadically before but with variable degrees of success [9,33,34]. Our principal goals were to achieve primary closure of the wounds without tension and with acceptable aesthetic outcome. Furthermore, we also aimed at providing primary reconstruction of missing structures as well as granting a second reinforcing layer over the dura in the pyopagus twin and chest wall in thoraco-omphalopagus variety. Although tissue expansion entails two procedures, it is not without many advantages. Other than the obvious pros in providing soft tissue coverage with ready-made flaps upon separation that are predictable and reliable, it allowed the anesthetic team to rehearse prior to the actual separation and for the pediatric surgeons to perform their abdominal laparoscopy and other essential investigations under anesthesia. Furthermore, having ready-to-use flaps upon separation obviates the unnecessary extra blood loss when dissecting flaps and shortens the reconstruction period, thus decreasing the overall operative time at separation.

Tissue expanders have been reported to be placed either in the subcutaneous plane [6,8,9,30,31,33] or intraabdominally [16,34]. They however, were not without complications and were reported to be associated with infection and skin necrosis with resultant skin loss in those inserted subcutaneously and bowel perforation and obstruction in those inserted intraabdominally [16,34]. Unlike Filler’s and Zuker et al., preference of the intraperitoneal position [16,34] and in accordance with others [6,8,9,30,31,33], tissue expanders were inserted in the subcutaneous plane in our twins. We did not encounter any complications in the two sets of twins. This could be explained in part by avoiding the risk factors that predisposed to failure of expansion in the previous reports. The incision for insertion was put as remote as possible and radial to the longitudinal axis of the expander to eliminate the risk of exposure and extrusion. An internal port was used to prevent the higher risk of infection if external ports were used and also, to avoid pulling or damaging by the babies. These internal ports were fixed at a far distance to avoid the overlap as we relied on the overexpansion technique to gain more extra tissues with the minimal number of expanders used. The fact that only one expander was used decreased the area of undermining, pocket size and ultimately reduced the risk of infection. This could not have been achievable unless the expander was placed as close as possible to the advancing skin edge, hence the site of placement along the fusion area. Furthermore, using a single expander and choosing the exact site along the fusion area was based on being devoid from pressure points while the twins were asleep or turning around in bed to facilitate their nursing during the expansion phase. Additionally, the rate of the expansion process was slow which provided a safer technique and avoided skin necrosis, underlying
deformity and movement restriction in rapid expansion. It also gave a chance for more maturation of the expanded skin flaps and the pseudocapsule limiting the rate of anticipated retraction following its delivery. Synder [38] advocated the advance the skin flaps as soon as possible, since it will resume its original size in a short time. That is why we adopted the slow expansion technique and waited for two extra weeks after full expansion and prior to attempting the separation. Also, the expander was delivered last in the thoraco-omphalopagus twin at the time of separation and immediately prior to reconstruction and closure. In the pyopagus twin the expander had to be delivered first and the expansion was maintained throughout the separation procedure by inserting gauze underneath the expanded skin.

Another consideration should be given to the expanded area. We preferred to expand the bridging area with a single large volume expander similar to Hockley et al. [7]. This was found to yield expanded skin flaps for both twins. To the contrary, most of the previous reports expanded the surrounding areas to the shared region with insertion of multiple expanders with its subsequent complications [6,8,9,16,23,28,30-34]. In a recent report by Losee et al. [31] in separating an ischiopagus twin, they inserted nine expanders circumferentially around the shared area. This was followed by multiple pocket infections with its removal after one week. They repeated the expansion three months later placing four expanders but still circumferential. Again, this resulted in circumferential compressive forces with expansion and development of restrictive lung disease with ultimate abruption of the procedure to save the life of the infants. They recommended placing fewer but larger expanders along their shared surface area only to allow for free chest and abdomen expansion. This is in agreement with the policy adopted in our cases. It is worth noting that one of the great advantages of using a single expander is to avoid all the hazards of frequent turning and repositioning of the twins intraoperatively prior to completing their separation.

The CSF leak has been reported to occur in 37.5% of separated pyopagi with conjoined cords [27,29]. In our twins, the technique of reinforcing dural closure by a second layer of pseudocapsule flap proved successful in preventing the CSF leak. This is of extreme importance in ensuring adequate wound healing especially in the presence of adjunctive gastrointestinal surgery to reduce the risk of sepsis. Recent reports in the separation of pyopagus twins have referred to the use of the pseudocapsule as a graft [7] or a flap [6,28] in providing an autologous layer to achieve watertight dural closure especially when primary dural closure was not possible. Adopting the same principle, this vascularized pseudocapsule flap was also used to reinforce chest wall closure over the synthetic patch in the thoraco-omphalopagus twin with rib cage defects. Over added coverage was achieved by the expanded skin flap that ultimately gave reasonable layered closure with favorable outcome. Although Al Rabeah in 2006 [5] recommended extreme selectivity in the use of tissue expanders and prosthetic meshes to avoid the potential risk of infection and unnecessary delay and cost, we believe that the use of expanded skin for coverage may be the optimal method at present for its unprecedented advantages when properly employed.

In conclusion, the primary repair of the developed wounds after separation of conjoined twins is problematic. Applying the proper expansion technique while using a single tissue expander provided abundance increase in the shared area, including skin and pseudocapsule, allowing for a solid two layer closure. It also obviates the complications associated with the use of traditional methods of wound closure and gives good cosmetic results. Although tissue expansion is a two-stage procedure, it allows the anesthetic team to rehearse prior to the actual separation and for the pediatric surgeons to perform their abdominal laparoscopy and other essential pre-separation investigations under general anesthesia. Furthermore, it prevented the unnecessary extra blood loss and decreased the overall operative time during the separation stage.

REFERENCES