Ultimate pediatric multidisciplinary team challenge

A case report of Conjoined Twin(PYGOPAGUS) separation

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Abstract
We report successful separation of PYGOPAGUS twins. Meticulous planning and multidisciplinary approach are the core aspects in the management of Pygopagus conjoined twins. Early elective separation versus delayed separation of the conjoined twins improves the survival from 40% to 80%. Preoperative assessment of sickle cell disease, cross-circulation and organ sharing between twins proved valuable for planning sedation and analgesia and allowing detailed scripting of the complex anesthetic induction sequence, airway management, complex neurological monitoring and post operative intensive care related issues. Fusion of lower back, spinal cord, nerve roots, and rectal and genitourinary tract was a major surgical challenge related to potentially adverse neurological outcomes, bladder/bowel dysfunction and intact long term survival. Post operative critical care issues such as pain, prone ventilation, sickle cell sequelae, cerebrospinal fluid leak and meningitis, wound infection and colostomy care, nutrition, and other potential complications are discussed in detail in this report.

We emphasize the role of pediatric intensive care, preoperative planning with multidisciplinary team, and staged surgical intervention and a good postoperative care in the successful management of complicated variants.

Introduction
Conjoined twins are monozygotic, monoamniotic and monochorionic. The incidence is 1 in every 200 twin pregnancies, varying from 1 in 50,000 to 1 in 100,000 pregnancies[1]. Female conjoined twins are more common and account for 70-90% of cases[1]. There is a reported racial predilection, with increased incidence in Africa and South-East Asia[1].

Conjoined twins are typically classified by the point at which their bodies are joined, with the suffix “pagus”, which is Greek for “that which is fixed”. The commonest types are: Thoracopagus—40% of cases where two bodies are fused from the upper thorax to the lower abdomen (heart is usually always involved). Omphalopagus—33% of cases are fused at the lower chest (heart is not usually involved). Twins share the liver, other parts of the gastrointestinal tract and diaphragm. Pygopagus – 17% of cases; twins are fused at the spine, rectum, genitourinary tract. Criciopagus – Constitutes 2% of cases. Fused skulls, but separate bodies. Ischiopagus: Anterior union of the lower half of the body (joined at the pelvis) occurring in about 6% of all conjoined twins. Parasitic Twins: Rare forms of conjoined twins, asymmetrical conjoined twins, one twin being small, less formed and dependent upon the other. With this background we report our experience of preoperative evaluation, multidisciplinary planning, parenteral sedation for series of diagnostic procedures and postoperative management in the Pediatric intensive care unit.
Case:

8 months old, female conjoined twins born out of a nonconsanguinous marriage were delivered through normal vaginal route at a hospital in Nigeria on 28th Aug 2012. Combined Birth weight was 4.5 kg. Antenatal Ultrasound (USG) was suggestive of twin pregnancy, there was no prior history of twins in the family. Twins were kept in the hospital for about 8 months before being referred for surgical separation. They did not have any major health related issues and were growing well with intact neurological status and bladder bowel function appropriate for age. Parents were informed regarding risk of potential loss of life of one of the twins during surgical separation by doctors in Nigeria. That led the parents seek alternative advanced surgical centres to get the best possible outcomes. Parents chose to come to our centre located in central Delhi; well equipped with the multidisciplinary pediatric medical and surgical teams, operating tertiary care facility and a state of the art pediatric intensive care.

At the time of admission, combined weight of conjoined twins was 13.8 kg. General and systemic examination was within normal limits. Twins were fused at the level of sacrum, with both children aligned in opposite directions, lower limb movements being normal. They shared common genitalia with single and dysplastic sacrum, spina bifida at level L5 and S1 vertebrae.

Medical team was appointed by Medical administration (CEO), who took the lead in supervising the overall planning and execution by the multidisciplinary team members, including planning of counseling by the interpreters and liaison with the family regarding the financial and other practical logistics. Team consisted of pediatric surgeon, pediatric intensivist as the main surgical and medical team leaders along with key superspecialists such as plastic surgeons, neurosurgeons, anesthesiologists, urologists, gastro Intestinal surgeons, gynecologists, vascular surgeons, radiologists and senior pediatricians.

Children were investigated in detail. Routine investigations included complete hemogram, liver and kidney functions, coagulation profile, type and screening, sickle cell trait, chest radiograph, echocardiogram, USG (ultrasound) abdomen, CT angiography, MRI lumbo-sacral spine and cystourethrogram. Twins were positive for sickle cell trait. To test for cross circulation between the twins an atropine test was done in the pediatric intensive care unit before exposing the twins to parenteral sedation required for imaging studies (MRI and CT scan). The atropine test was negative and revealed no major changes in the heart rate of the second twin when atropine was given to the first twin. To avoid errors from the very first day twins were marked as first and second twin and later on when they underwent surgical procedures, they were also color coded as pink and blue.

Various imaging studies concluded that twins had fusion of lower conus medullaris at S1 syrinx, single filum terminale, bilateral thecal sacs fused at S1 and ending at S2. There was fusion of left internal iliac vein of right side twin with internal iliac vein of left side twin at the level of acetabulum, with minor shunting of blood from one twin to other and vice versa. Both twins had their respective abdominopelvic organs such as urinary bladder, urethra, both ovaries, uterus, possibly vagina, rectosigmoid colon and rectum. There was a single puborectalis sling with single anal canal below it. Both urethras were found converging in the midline and became imperceptible in their distal course. In the midline, the two rectums were separated by a thin hypointense septum, where as both pelvic bones appeared unremarkable.

Literature was reviewed in detail. Anticipated risks, complications and ethical issues were discussed amongst the team members as well as with the family. Major concerns were risk of post operative neurological deficit, incontinence (bladder and bowel dysfunction), potential CSF leak, wound infections and meningitis after reconstruction surgery as well as the risk of mortality. Parents were counseled by the team through an interpreter. An option of not going for surgery was also discussed, however, in view of potential later morbidity and poor prognosis, family consented to go for separation surgery. After informed consent was obtained, all
major and minor risks explained, surgery was planned in 3 stages. All stages of surgery were rehearsed with
detailed documentation of designation of team leaders and respective members, their individual roles and all
major and minor equipment was secured. For stage 2 a neuro stimulator monitoring technology was also
secured to ensure proper separation of nerve roots in the sacral area which were anticipated to be fused,
entangled or crossing over.

Overall Plan

Stage 1: Placement of tissue expanders to obtain enough skin for wound coverage.

Stage 2: Separation of twins with reconstruction and diverting colostomy

Stage 3: Colostomy closure

As a part of stage 1, tissue expanders were placed in the pelvic regions to obtain expansion of skin and
subcutaneous tissues. Babies tolerated anesthesia and the entire procedure well and were subsequently
discharged. After 2 months, children were readmitted for the 2nd stage of surgery. Basic work up was repeated.
In view of normal hemoglobin with sickle cell trait, fresh blood was arranged to replace blood loss as per the
recommendations of transfusion specialist to avoid any acidosis leading to risk of sickle crisis and ischemia of
skin or other tissues. Twins were shifted to PICU night before surgery, central venous (subclavian) and arterial (radial) lines were accessed in each twin. Bowel was prepared and maintenance intravenous fluid was started.
Vancomycin, ceftriaxone and metronidazole were added for perioperative antimicrobial prophylaxis in view of
bowel, urinary and spinal dural surgery in the pelvic area. Twins were kept nil by mouth after 3 am on day of
surgery and shifted to operating room at 6am.

First incision was given by plastic surgeons, flaps were raised in prone and supine position, separated till
sacrum, significant blood loss and hypothermia were anticipated. Sacral vertebrae were divided in the
midline, neurosurgeons separated the nerve roots using two microscopes and two neuro-stimulators to ensure
muscle innervation of lower extremities of respective twins, cord/filum terminale was divided and dura was
repaired.

Pediatric Surgeon stood at the foot end, babies were brought to the edge of table with legs wide separated
supported with padding. Multiple stay sutures were taken to expose the perineum. Vagino-urethroplasty was
done for each twin before separation.

Anoplasty was planned after separation when the sacral flap would be in place. Sphincters and perineal body
were reconstructed. Right transverse colostomy was done in supine position.

Twins were separated successfully and further repair was carried out followed by closure of wounds in two
separate operating rooms with two surgical and anesthesia teams. The total duration of surgery was 18 hrs with
estimated blood loss of around 750 ml.

Consultant pediatric intensivists were present throughout the entire duration of surgery to ensure assistance in
medical management. Both twins were shifted to pediatric intensive care unit for elective ventilatory support
where they were placed in prone position which was a challenge as children had a big wound at the back covered
by a skin flap and colostomy in the front and at all cost pressure necrosis and infection had to be avoided. In the
immediate post operative period, we found the hand made soft rings made of sterile gauze and cotton very
useful which were so placed that they formed the groove for the colostomy wound and thus prevented any direct
pressure on it. However once children were successfully extubated after 48 hrs their preoperative habit of lying
in the prone position due to placement of tissue expanders came in handy to manage the postoperative period
successfully. One of the twins had mild metabolic acidosis postoperatively which resolved in the next 24 hours.
Pain was managed with multimodal approach, pharmacological agents used were Fentanyl and Paracetamol.
Sickle cell trait did not pose any significant problems as fresh blood was transfused, acidosis and hypoxia was avoided at all times.

Postoperatively Twin 1 had minimal weakness in right lower limb and Twin 2 had a minor CSF leakage which gradually stopped after 5 days. Presently after a period of 3 months of follow up, both the twins are without any neurological deficit and are continent for stools and urine. They have started walking with support.

Discussion

Management of conjoined twins remains a challenging task. It not only involves risks related to medical issues but ethical justification of risk taking. It is therefore a joint decision of the medical team and the family to go ahead with the separation or not, as the survival is fifty percent with possible significant morbidities such as limb paralysis, bladder and bowel dysfunction. It requires a multidisciplinary approach, thoughtful and meticulous planning to take care of all the aspects. This comprises extensive medical work-up on patients, multiple meetings and discussions with all the involved specialties and supporting staff, involvement of parents, psychosocial counseling of parents and rehearsal of the planned surgical procedure. The rationale for deferring surgery should include single heart, major communicating hearts or major anomalies.

Estimation of cross circulation is very important for two reasons, first, to calculate drug doses right from the beginning as drugs administered to one twin may have unexpected effects on the other while performing various diagnostic procedures and second, to calculate the percentage of cardiac output shared by the twins especially before surgery as one twin may be dependent on the other circulation for survival. Though circulatory admixing is more common in the thoracopagus and craniopagus twins than in other types, still, a careful angiographic or radio isotopic imaging of the cross-circulation is recommended for all conjoined twins.

The methods to ascertain the cross circulation in the conjoined twins are bedside atropine test (which we used in our case), injecting Tc-99m microcolloidal human serum albumin (HSA), Tc-99m HIDA, indigo carmine and the examination of its excretion in urine of the other twin. Szmuk et al were first to describe the role of bispectral index monitor for detection of cross-circulation in complex cyanotic heart disease in Thoracopagus conjoined twins.

Intraoperative period is unusual mainly due to abnormal position and presence of two patients on the same table simultaneously, in addition to involvement of multiple specialties along with duplication of medical personnel (for each twin) and equipment to handle them independently in the same operation theatre. Performing mock drills using mannequin specifically for change of posture from supine to prone and vice versa, shifting the patient head to the foot end of the table are really helpful. Besides that issues related to prolonged surgery, massive blood loss and hypothermia need to be anticipated and managed.

Postoperative period in an intensive care unit is crucial for stabilization of the separated infants. Elective ventilation and paralysis in the immediate postoperative period for 24-48hrs are preferable to achieve hemodynamic stability and fluid and electrolyte balance. Perioperative antibiotics and strict infectious precaution are recommended as sepsis is the major contributory factor in determining the outcome of the separated infants. The other important issue is a large skin defect as flaps and graft has been utilized in closure. To prevent pressure related necrosis and to maintain vitality of soft tissue various techniques are employed like nursing in prone posture with frequent turning, supportive gel padding and immobilization when needed. Early nutritional rehabilitation had a key role and it should be instituted early as it augments wound healing and prevents infectious complications.

The incidence of CSF leak in post operative period in pygopagus twins with conjoined cords who survive surgery is about 40%. In our case one of the twins also had CSF leak which gradually subsided after 5 days.
however in the previous case reports children required re-operation once or twice to undertake dural repair.

Till date 19 operative cases of pygopagus conjoined twins have been reported in the literature and 8 of these, had some degree of neural fusion as in our case. Elective separation may take place as early as 2-4 months of age with survival rate of 80-90% in most series as compared to emergent surgery where it falls to 30-50%. The obvious advantages of separation after 2-4 months of age (as opposed to right at birth) are diminished risk of anesthesia, ability to evaluate anatomy and congenital anomalies and ensure better wound coverage by placing pre separation tissue expanders.

To conclude, the successful separation of twins emphasizes the need for team work and planning every minute detail; from preoperative evaluation of cross-circulation and organ sharing, long surgery with blood loss and massive fluid shifts and post operative care in the pediatric intensive care unit (see figures 1 to 15). Finally, the sight of two separated smiling babies lying on either side of the mother without any obvious neurological deficit is very rewarding and worth all efforts.

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Source of Funding: None

References
Table: Pygopagus twins with union involving neural structures

<table>
<thead>
<tr>
<th>Reference</th>
<th>Year of surgery</th>
<th>Sex</th>
<th>Shared neural elements</th>
<th>Other anomalies</th>
<th>Timing of operation</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>Luce 1956</td>
<td>Male</td>
<td>Two sensory roots</td>
<td>Single shared anus</td>
<td>Elective, 17 days old</td>
<td>Unknown</td>
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<tr>
<td>Gupta 1962</td>
<td>Female</td>
<td>Lower spinal cord, meninges and caudaequin a were common</td>
<td>Single shared anus; cardiac anomaly in “B”</td>
<td>Emergency, 5 months old</td>
<td>“A” normal at 5 years. “B” died intra-op</td>
<td></td>
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<tr>
<td>Votteler 1981</td>
<td>Female</td>
<td>Spinal cord in continuity</td>
<td>Single shared anus</td>
<td>Elective, 2 months old</td>
<td>Normal autonomic &amp;motor function</td>
<td></td>
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<tr>
<td>Kalo 1985</td>
<td>Female</td>
<td>Caudaequina partially adhered</td>
<td>Single shared anus; brain death in “B”</td>
<td>Emergency, 6 days old</td>
<td>“A” unknown “B” died 35 days post-operatively</td>
<td></td>
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<tr>
<td>Fowler 1994</td>
<td>Female</td>
<td>Shared distal cord</td>
<td>“A” patent rectum; no uterus/vagina; one kidney. “B” multiple: imperforate fistula; absent kidneys</td>
<td>Elective, 8 months old</td>
<td>“A” neurogenic bladder incontinent of stool, positional scoliosis. “B” died day 3</td>
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<td>Hockly 2001</td>
<td>Female</td>
<td>Fused lower spinal cords</td>
<td>B” no communication with anus</td>
<td>Elective, 3 months old</td>
<td>Survived; neurologically intact</td>
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<td>Fieggen 2003</td>
<td>Female</td>
<td>Distally fused spinal cords</td>
<td>Single shared anus</td>
<td>Elective, 8 months old</td>
<td>Survived; neurologically intact; CSF leak</td>
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<tr>
<td>Badr 2009</td>
<td>Male</td>
<td>Fused lower spinal cords</td>
<td>Single shared anus,</td>
<td>Elective, 8 months old</td>
<td>Survived; neurologically intact</td>
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<tr>
<td>Current Report 2013</td>
<td>Female</td>
<td>Fused lower Spinal cord</td>
<td>Single anus, Urethral opening</td>
<td>Elective 8 months old</td>
<td>Survived; Neurologically intact</td>
<td></td>
</tr>
</tbody>
</table>
PYGOPAGUS Twins separation

Fig 1. PYGOPAGUS conjoined lower back

Fig 2. Common genito urinary and anal openings due to fusion at the distal ends of rectum, vagina and urethra

Fig 3a common genitourinary opening and inferiorly common anal opening

Fig 3b shows a schematic diagram of fusion of distal ends of urethra and vagina and two rectums fused at distal common anal opening
Fig 4. CT reconstructed images of Bony fusion at lower back showing common sacrum

Fig 5. Lower end of spine and spinal cord showing fusion on MRI

Fig 6. MRI image of lower spine showing fusion of the spinal cord and filum terminale at the level of sacrum

Fig 7. CT angiography showed a common iliac vein crossing over(filling up) to other twin at the level of acetabulum

Fig 8. Two teams of anesthesia handling the twins for anesthetic induction

Fig 9. Surgical field with open dura showing distal end of Spinal cord and nerve roots fused and criss crossing, being separated by the neurosurgeon under magnification by microscope using nerve stimulator monitoring of distal muscular innervations of leg muscles of each twin.
**Fig 10.** Twin after separation being managed in prone position in the PICU

**Fig 11.** Separated Twin on 2nd postoperative day after elective ventilation for 36 hours just before extubation

**Fig 12.** Happy parents with the separated twin just after extubation receiving nebulisation

**Fig 13.** Separated twins doing well in Pediatric ICU 3rd postoperative day