

Parapagus conjoined twins: Complicated anatomy precludes separation

Arbinder K. Singhal, Gautam S. Agarwal, Shilpa Sharma, Arun K. Gupta*,
Devendra K. Gupta

Departments of Pediatric Surgery and *Radiodiagnosis, All India Institute of Medical Sciences, New Delhi, India

Correspondence: Dr. D. K. Gupta, Department of Pediatric Surgery, All India Institute of Medical Sciences, New Delhi - 110 029, India. E-mail: profdkgupta@gmail.com

ABSTRACT

A parapagus set of male conjoined twins was brought to our institution at 12 h after birth. An extensive sharing of the abdominal viscera (single liver, hindgut), abdominal aorta, pelvis (single rectum and anus), genitalia (one set) and vertebral column was found. The surgical separation was not considered due to medical and ethical issues.

KEY WORDS: Parapagus, conjoint twins

INTRODUCTION

Conjoined twins occur in one in 50-100,000 live births and are now thought to result from the fission of the notochordal anlage; followed by subsequent fusion of the embryos.^[1] Parapagus twins joined anterolaterally result from two nearly parallel notochords in close proximity. This anomaly represents less than 0.5% of all reported cases of conjoined twins.^[2] We present one such surviving set of conjoined twins.

MATERIALS AND METHODS

A set of male conjoined twins, born to a third gravida mother by vaginal delivery, presented to our institution at 12 h after birth. The mother had limited antenatal care, and no antenatal ultrasonogram was done. She had presented to a nearby hospital in advanced labor, and the delivery had been completed vaginally. The combined weight of the conjoined twins at the time of admission was 3.020 kg.

Clinical examination revealed two heads, two thoraces with side-to-side fusion in the lower half, four upper limbs, one abdomen, single set of genitalia and anus and two neurologically independent lower limbs [Figure 1]. At presentation in the emergency room, the right twin was cyanosed and hypothermic with poor activity. The line of demarcation between the cyanosed right twin and the pink left twin was in the midline in the thorax and the upper abdomen, while both the lower extremities

were cyanosed. The right twin was intubated and required ventilation for 24 h; after that, both the babies remained stable.

Babygram showed two separate vertebral columns with scoliosis and separate hemisacra, which joined distally [Figure 2]. Echocardiogram, ultrasonogram with doppler and contrast enhanced computed tomographic (CECT) scan revealed two structurally normal hearts with liver wedged in between; two separate thoracic aortas; two separate inferior vena cavae with ipsilateral drainage of hepatic veins; a central large liver with a single gall bladder; single spleen on the left side; and two normal kidneys, one in each flank. There were four lungs visualized; however, the medially placed lung of each baby was hypoplastic.

A magnetic resonance scan (MR) was done to evaluate the vascular anatomy and the spinal cords. The spinal cords were normal and only the dural sacs had joined in the sacral area. The two thoracic aortas joined in the abdominal region for only a short distance and then divided again into iliac arteries. This union of aortas was peculiar and led to distal cross circulation, explaining the presence of cyanosis in both the lower limbs at presentation.

RESULTS

The present set of twins was thus symmetrical, and the surgical separation was not considered due to medical

and ethical reasons. The separation would have resulted in division of common vital organs, maybe at the cost of losing one of the babies. The key issue that made us reject separation was sharing of the part of the vertebral column with peculiar innervation which was separate to either lower half of the body. Separation would have also entailed division of the single bony pelvis leading to gross deformity necessitating lifelong rehabilitation and family support. Furthermore, there was only one set of external genitalia, lower urinary tract and lower gastrointestinal tract, and their division would have led to gross morbidity. Parents did not give consent for separation in view of expected morbidity and high risk of mortality.

DISCUSSION

Literature abounds with confusing terminologies and classifications of conjoined twins, and to end all this, a standardized terminology was put forward by Spencer.^[3] According to this simplified classification, which is universally accepted now, the present case was classified as *dithoracic parapagus* with further specification as *tetrabrachius, bipedus* conjoined twin.



Figure 1: Clinical photograph of the twins



Figure 2: (a) Infantogram showing the skeletal anatomy (b) Magnetic resonance angiography showing union of abdominal aortas for a short distance

A simple clinical examination is sufficient to identify the type of twinning. Investigative work-up aimed at unraveling the anatomy and providing the road map for possible surgical management is done with a three-dimensional CECT and/or MRI scan. In parapagus twins, cardiac malformations are quite frequent; but in the present set, the MRI scan and echocardiogram showed normal cardiac anatomy.^[4]

The clinicians, however expert, are challenged by the day-to-day management of the twins with their individual pathology and anatomy. If a surgical intervention is contemplated, then it may be associated with complex and occasionally insurmountable legal and ethical difficulties.^[5] While in others, it is clearly not technically possible to attempt separation. Although life as a conjoined twin would seem intolerable, there are numerous instances of quality survival in many sets of non-separated twins.^[6,7]

Studies of long-surviving twins have identified that they develop distinct personalities and express different levels of behavior and intelligence.^[5,6] Despite this individualism, many long-living conjoined twins – even when faced with death – refused the option of separation. Conjoined twins are a different conceptual organism; and provided the twins are healthy and surviving, a very conservative medical approach seems to be indicated.^[6,7] Ethics impose on the clinician a need to act in the best interest of the child, but in conjoined twins we have competing interests. Barilan observes that ‘conjoinment challenges our sense of selfhood and identity.’^[7] This concept tends to drive the clinicians as they examine the possibility of separating the twins. In some cases, it is possible one twin will die as a result. The right and wrong of this approach can provoke widespread ethical, religious and media debate. The sanctity of life and the quality of life of the twins should be the central foci. Only then we can reach a consensus regarding the management of such unique cases. Given these issues, a common consensus was reached between clinicians and parents to leave the kids as such.

Presented as a poster

at the 29th annual conference of Indian Association of Pediatric Surgeons at Hyderabad, India, November, 2003.

REFERENCES

1. Spencer R. Theoretical and analytical embryology of conjoined twins: I. Embryogenesis. *Clin Anat* 2000;13:36-53.
2. O'Neill JA Jr, Holcomb GW 3rd, Schnauffer L, Templeton JM Jr, Bishop HC, Ross AJ 3rd, et al. Surgical Experience with Thirteen Conjoined Twins. *Ann Surg* 1988;208:299-312.
3. Spencer R. Anatomic Description of Conjoined Twins: A Plea for Standardized Terminology. *J Pediatr Surg* 1996;31:941-4.
4. Tansel T, Yazicioglu F. Cardiac and other malformations in

- parapagus twins. Arch Gynecol Obstet 2004;269:211-3.
5. Atkinson L. Ethics and conjoined twins. Childs Nerv Syst 2004;20:504-7.
 6. Raffensperger J. A Philosophical Approach to conjoined Twins. Pediatr Surg Int 1997;12:249-55.
 7. Barilan YM. One or two: An examination of the recent case of the conjoined twins from Malta. J Med Philos 2003;28:27-44.