

# Epigastric Heteropagus Twin: Our Experiences of two cases with review of literature

S Wadhera, S Singh, B Lahoti, M Rajkumar

## Citation

S Wadhera, S Singh, B Lahoti, M Rajkumar. *Epigastric Heteropagus Twin: Our Experiences of two cases with review of literature*. The Internet Journal of Surgery. 2007 Volume 14 Number 1.

## Abstract

Conjoined twins occur in approximately one in 50,000 births. Considerable debate persists over the development of conjoined twins; some have speculated whether they develop as a result of fusion of embryos at specific embryological sites or as a failure of fission. Asymmetric and parasitic conjoined twins are rarer anomalies of monochorionic monoamniotic twins, consisting of an incomplete twin attached to the fully developed body of the co-twin.

## INTRODUCTION

Conjoined twins occur in approximately one in 50,000 births (1). Asymmetric and parasitic conjoined twins are rarer anomalies of monochorionic monoamniotic twins, consisting of an incomplete twin attached to the fully developed body of the co-twin (2). The term Heteropagus Twin was given by Potter & Craig (3), while Willis termed this condition as Parasitic United Twins (4). Omphalopagus refers to conjoined twins joined at the level of the umbilicus (5).

## CASE REPORT 1

A two day full-term normal delivery male, total weight 3.8 kg, had an omphalocele with intact membranes (autosite) and a parasite with a pelvis, well formed lower limbs without movements, one rudimentary upper limb, normally developed external genitalia and an imperforate anus (Fig. 1). Hematological and biochemistry work-up was within normal limits. X-ray showed developed bones in the lower limbs of the parasite, the host did not show any anomaly. Ultrasonography of the host showed the omphalocele sac with liver. The truncus showed a single fused pelvic kidney, the connecting bridge showed bowel-like cystic tissue. Echocardiography showed a large ventricular septal defect. Elective surgery was done with separation of the truncus from the host (Fig. 2). The joint part showed two smaller vessels, the truncus had unused intestines in the host's omphalocele and an atretic colon. The rest of the content of the omphalocele was the liver of the host. The parasitic truncus had one large pelvic kidney, ureter and bladder. The weight of the truncus was 650g. Postoperative recovery was uneventful and the baby was discharged on the 9<sup>th</sup> day. The

child was doing well in follow-up.

## Figure 1

Figure 1



## Figure 2

Figure 2



## CASE REPORT 2

A full-term neonate presented on the first postnatal day with an omphalocele with intact membranes. The host was feeding well and passing meconium. Combined weight was 3.2 kg. The parasite had a partly formed face with hair at the site of the head, rudimentary upper limbs and a palpable vertebral column (Fig. 3). CT and MRI showed the parasite sharing a common liver and a part of small bowel with the autosite. Cardiac Doppler showed mild tricuspid regurgitation (TR) with patent ductus arteriosus (PDA) with a left-to-right shunt. The parasite was separated successfully on the 10<sup>th</sup> postnatal day, the liver was preserved in the autosite, and a blind ending loop of jejunum (Fig. 4) which was common was excised with jejuno-jejunal anastomosis. The truncus weighed 400g. The baby succumbed on the 1<sup>st</sup> postoperative day as a result of cardiac failure.

**Figure 3**

Figure 3



**Figure 4**

Figure 4



## DISCUSSION

There is considerable debate over the development of conjoined twins; some have speculated whether they develop as a result of fusion of embryos or failure of fission.

One theory proposes that conjoined twins develop from a single fertilized ovum and result from failure of division of the embryonic disk until after day 13 from conception. Seventy percent of conjoined twins are female, and 40% are stillborn (6).

Another theory states that conjoined twins are categorized based on the region of fusion: Fusion at neural pores (Craniopagus, Pygopagus) or at the oropharyngeal membrane (Cephalopagus). Omphalopagus and Thoracopagus are due to fusion at the primordial location of diaphragm and heart, respectively, as both diaphragm and heart lie outside the rostral edge of the embryonic disc (7).

Some authors have suggested that this anomaly results from an ischemic atrophy of the body structure of monozygotic conjoined twins at an early gestational age leading to selective atrophy of the cranial part of one of the monozygous twins (8). This may lead to reversal of the circulation of the deformed twin, i.e. deoxygenated blood is supplied by the umbilical artery of the normal twin and this may lead to severe malformation. In a Parasitic Omphalopagus there is associated congenital heart disease in the autosite of varying degree (7).

## CONCLUSION

As the controversy over the development of conjoint twins persists, we think that this rare phenomenon which is often unreported in developing countries is a result of a single

fertilized ovum and results from failure of division of the embryonic disk until after day 13 from conception.

### **References**

1. O'Neill JA, Jr, Holcomb GW, 3rd, Schnauffer L, Templeton JM, Jr, Bishop HC, Ross AJ, 3rd, Duckett JW, Norwood WI, Ziegler MM, Koop CE. Surgical experience with thirteen conjoined twins. *Ann Surg* 1988; 208(3): 299-312.
2. Gupta DK, Lall A, Bajpai M. Epigastric heteropagus twins - a report of four cases. *Pediatr Surg Int* 2001; 17(5-6): 481-2.
3. Potter EL, Craig JM. *Pathology of Fetus and Newborn* (ed. 3), Chicago IC Year Book 1975; 220-237.
4. Willis RA. *Borderland of embryology and pathology*. Austin, TX, Butterworth. 1962.
5. De Ugarte DA, Boechat MI, Shaw WW, Laks H, Williams H, Atkinson JB. Parasitic omphalopagus complicated by omphalocele and congenital heart disease. *J Pediatr Surg* 2002; 37(9): 1357-8.
6. Ahmet Gul, Halil Aslan, and Yavuz Ceylan. Prenatal diagnosis of pygopagus tetrapus parasitic twin: case report. *BMC Pregnancy Childbirth*. 2004; 4: 13.
7. Keith W Ashcraft. *Pediatric Surgery*, 3rd Edition, 1040-1045.
8. Raynal P, Petit T, Ravasse P, Herlicoviez M. Un cas rare de jumeaux heteropages epigastriques. *Journal de gynecologie obstetrique et de biologie de la reproduction* 2001; 30: 65-69.

**Author Information**

**Sushant Wadhera, MS(pg)**

Department of Surgery, M.Y. Hospital

**Sangram Singh, Mch Paed Surg**

Department of Surgery, M.Y. Hospital

**Brajesh K. Lahoti, Mch Paed Surg**

Department of Surgery, M.Y. Hospital

**Mathur Rajkumar, M.S., F.I.C.S, F.A.I.S.**

Department of Surgery, M.Y. Hospital