

## Undiagnosed xiphopagus twins: a perinatal malady

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### Abstract

Conjoined twins are a very rare entity. It is associated with poor survival rate in the presence of vital organ sharing. The entity can be diagnosed as early as the first trimester. A conjoined twin diagnosed late in labor is a malady with high perinatal mortality and maternal morbidity. We present one such case of xiphopagus twins. The management of a case diagnosed late in labor can be very challenging. Such obstetric challenges can be avoided by a meticulous early scan with a high index of suspicion, especially in the absence of separating membrane while scanning multiple pregnancies.

### Introduction

Conjoint twins are very rare. The incidence is about 1 in 60,000 to 1 in 100,000 deliveries.<sup>1</sup> In a large multicenter worldwide research that studied the largest sample of conjoined twins, the total prevalence was 1.47 per 100,000 births.<sup>2</sup> The condition can be diagnosed early in labor is an unfortunate malady associated with very high perinatal mortality and maternal morbidity. We report one such case of xiphopagus twins diagnosed in advanced labor. Such a malady can be easily prevented if a checklist is followed while performing an ultrasound for a multiple pregnancy.

### Case Report

A 32-year-old, nulliparous woman with one previous abortion was admitted at 40 weeks of pregnancy with labor pains. She had had a non-consanguineous marriage two years earlier. She was admitted to the labor room at term with pains and draining for half hour. She had been already diagnosed to have a twin pregnancy at 30 weeks of gestation by a general practitioner. There was no history of prior usage of contraceptive pills or ovulation inducing agents. There was no history of twinning

in the family.

At admission her blood pressure was 140/100 mm in the right arm supine position. She had mild anemia. There was mild pedal edema. Abdomen was over distended with multiple fetal parts. The first twin presented as vertex (left occipito-transverse). The second head was felt in the fundus of the uterus. Both fetal heart sounds were good. Uterus was acting mildly. The expected combined fetal weight was 5 kg. There was no polyhydramnios. On vaginal examination, the cervix was 3 cm dilated, fully effaced and vertex was at -3 station. Membranes were absent and clear liquid was draining. Pelvis was normal gynecoid.

The patient was monitored. An intravenous access was secured and she was sedated. Investigations revealed normal bleeding time, clotting time and platelet count. Blood urea and creatinine levels were within normal limits and there was no albuminuria. Her blood pressure and progress of labor was monitored. Uterine contractions were regular and optimal in intensity. Her cervical dilation and descent of the first vertex progressed satisfactorily with the partograph line to the left of the alert line. The fetal heart sounds were well preserved. Her diastolic blood pressure varied between 90-100 mm of Hg. The labor was uneventful till the second stage. Perineal infiltration was given and episiotomy was made. The head of the first twin delivered spontaneously followed by shoulders and upper limbs. The chest appeared till the lower part of sternum. Thereafter the labor came to a standstill. The umbilical cord was not felt. A cartilaginous stalk was felt from the lower end of the sternum. Traction on the delivered part did not cause any further descent. Neither could the limits of the stalk be made out from the vaginal approach. Unfortunately, by this time none of the fetal heart sounds could be clinically auscultated. Suspecting conjoined twins, she was prepared for abdominal delivery. Lower segment was opened; the second twin presenting as breech was held. There was a cartilaginous stalk connecting the twins from the xiphi-sternum to the mid-abdomen. The conjoined twins were carefully delivered intact from the uterus after reverse manipulation from the vagina aided by traction on the second twin from above. Both the fetuses were born dead. There was no extension of the uterine incision or post partum hemorrhage.

Xiphopagus twins, both female, with a combined weight of 5.4 kg were still born (Figure 1). There was a single placenta and single umbilical cord attached to the middle of the stalk. The cord revealed two arteries and one vein with a velamentous insertion (Figure 2). Postnatal skeletal survey showed a cartilaginous connection, normal thoraces and normal skeletal morphology (Figure 3). The relatives refused autopsy of the fetuses. The patient had

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a problematic post-operative period with pyrexia and wound infection. The wound was re-sutured after controlling infection with antibiotics as per the culture report. She was discharged from the hospital in good condition after 21 days.

### Discussion

Conjoint twinning occurs in monozygotic twinning when the embryo divides 13-14 days after fertilization. Xiphopagus twins are joined at the xiphoid process (part of the sternum) and usually linked only by cartilage and soft tissue. These twins share no vital organs but often have conjoined livers. This is a type of ventral union. Xiphopagus is terata anacata-didyma type of twins (Guttamacher's classification). Three percent of conjoined twins are xiphopagus. They are more common in females (3:1).<sup>3</sup>

Chang and Eng Bunker (1811-1874) were the first known xiphopagus twins with conjoined livers. Their autopsy had revealed a band of liver along with the xiphoid with a median bursa like sac. The synchondrosis had been stretched by these active twins to enable them to stand side by side.<sup>4</sup>

Harper *et al.*<sup>5</sup> (1980) studied 36 pairs of xiphopagus twins over 300 years. They observed that 69% were female, 10% were associated with polyhydramnios, 50% had cephalic-breech presentation, 36% had dystocia and the still birth rate was 19%. They further reported that 6% had only a skin bridge, 81% had conjoined liver, 56% had conjoined sterna cartilage, 17% had conjoined

diaphragm, 3% had conjoined genitor-urinary system and 33% had associated other malformations. Twenty-seven of these thirty-six pairs had been operated upon of which 18 sets and 7 singles survived. The neonatal complications and mortality increased as the number of organs shared increased.

Prognosis of these twins depends on the



Figure 1. The xiphopagus twins.



Figure 2. The velamentous insertion of the single umbilical cord.



Figure 3. X-ray of the twin showing normal skeletal survey and separate thoracic cages.

nature and extent of organ sharing and presence of other malformations. Hoyle<sup>6</sup> (1990) analyzed all attempts at surgical separation of conjoined twins reported in the literature until 1987. Surgical separation had been attempted on 167 occasions. The overall survival rate in this group of twins was 64%. The highest mortality occurred among thoracopagus (51%), craniopagus (48%), and omphalopagus (32%). Mortality was lower with ischiopagus (19%) and pygopagus (23%). He also noted a significant increase in mortality for emergency separation (70%) compared with elective procedures (20%).

In a recent study,<sup>7</sup> 36 twin pregnancies with conjoined twins seen over a period of 12 years in a single tertiary hospital were analyzed. Of these, 69.4% were thoracopagus and 5.6% were omphalopagus. Cardiac defects were the most frequent, present in 91.6% of twin pairs. Associated malformations were present in 61.8% of the cases. Twelve pregnancies with poor prognosis were terminated and the remaining cases were delivered by cesarean section. Five sets of twins underwent surgical separation and 6 children survived. Overall survival in these was only 8.3%.

Xiphopagus twins with only soft tissue bridge, as in our case, carry a very good prognosis after separation, with the earliest survivors operated way back in 1952.<sup>8</sup> Shukla and colleagues reported the case of a pair who was delivered by cesarean for prolonged labor.<sup>9</sup> In this case, also the diagnosis had been missed in the previous scans performed during pregnancy. One of the twins had multiple malformations and died after birth. In this xiphopagus pair, liver was shared. Emergency separation had been undertaken as it was threatening the survival of the live fetus.

Various techniques have been used to delineate the hepatobiliary and vascular sharing amongst the xiphopagus twins before planning separation surgery. Intravenous cholangiography has been used to delineate the biliary tree and gall bladder.<sup>10,11</sup> Upadhyay<sup>10</sup> used Iodine-I 31 rosebengal. Kling *et al.*<sup>11</sup> used umbilical vein angiography. Sequential scintiangiography technique using Tc-99m sulfur colloid was used by Margouleff *et al.*<sup>12</sup> for the delineation of the size, configuration, anatomic point of fusion, and quantity of cross circulation in xiphopagus conjoined twins.

In our case, it was extremely unfortunate that, in spite of two scans performed on two different occasions by two different observers in the second trimester, this problem was missed. The scan reports did not make any comment on the separating membranes. Presence of separating membrane should form an important checklist while scanning twins. Diamniotic placentation almost always rules out conjoined twins with the exception of 3 cases reported in literature.<sup>13-15</sup> The absence of

separating membrane indicates monoamniotic twins. In such cases detailed, patient and repeated scans may be necessary to rule out fixed relationship of fetus to each other and hence the possibility of conjoined twins.

The Canadian Task Force on Preventive Health Care has made recommendations for the best use of ultrasound in twin pregnancies based on the study by Morin and Lim.<sup>16</sup> The authors emphasize the need to adhere to these recommendations to reduce such preventable maladies.

Early diagnosis with scans keeping the checklist in mind can avert unwarranted challenge and dilemma to the obstetrician in the labor room, and morbidity and extreme pain and psychological anguish to the mother. Planned delivery in appropriate centers with expertise for separation and post-operative support once the extent of organ sharing is known can reduce the perinatal mortality and maternal morbidity.

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