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CONJOINED ISCHIOPAGUS TWINS*

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Conjoined twins are a rare abnormality. The frequency of the birth of conjoined twins is said to be in a ratio of from 1/25000-1/80000[1-3]. There are approximately 600 articles written on this subject, and from these it is understood that the life span of conjoined twins is between a few hours and 63 years[4-7]. Ischiopagus is seen in about 6% of all conjoined twins[6,8]. In this article a case of conjoined ischiopagus twins, the first to be surgically separated in Turkey is presented.

Case Report

The babies were delivered normally at term at a hospital in Gaziantep in Southeast Turkey after a normal gestational period. Three days after their birth the twins were transferred to the Department of Pediatric Surgery of Çukurova University Hospital in Adana.

On physical examination it was found that the axillary temperature of each baby was 35 °C, their pulse was 130/min and their combined weight was 4500 grams. At the point of combination an omphalocele, 6 cm in diameter, was noted. It was also observed that on each lateral side there were openings resembling a urogenital sinus. Apart from this, the first baby (Baby I) showed no other abnormality. The second baby (Baby II) was found to have Pierre Robin syndrome, a bilateral upper extremity abnormality, and hypoplasia of the external ear. Advanced cyanosis was also observed (Figure 1a-b). In contrast to Baby I, no respiratory sound was heard; but a pansystolic murmur in the heart, grade 4/6, was noted in Baby II. Results of laboratory examinations were as follows: hematocrit 55%, blood glucose concentration 40 mg/dl, Na 136 mEq/L, Cl^- 94 mEq/L, K^+ 3.8 mEq/L, BUN 26 mg/dl. Intravenous pyelography showed that the babies had four kidneys and two bladders. Each bladder was situated on either side of

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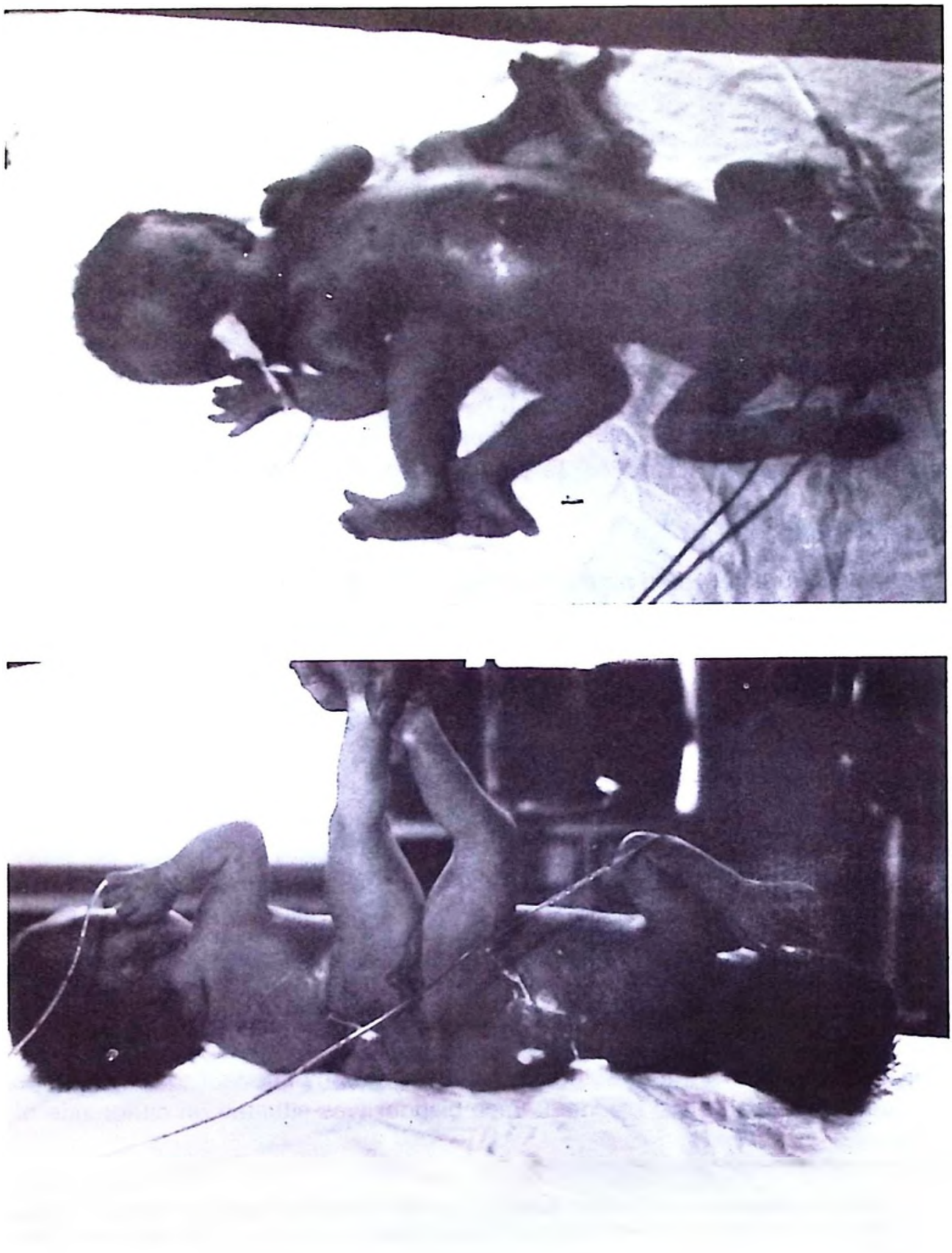

Figure 1

The anterior (a) and lateral views (b) of the twins.

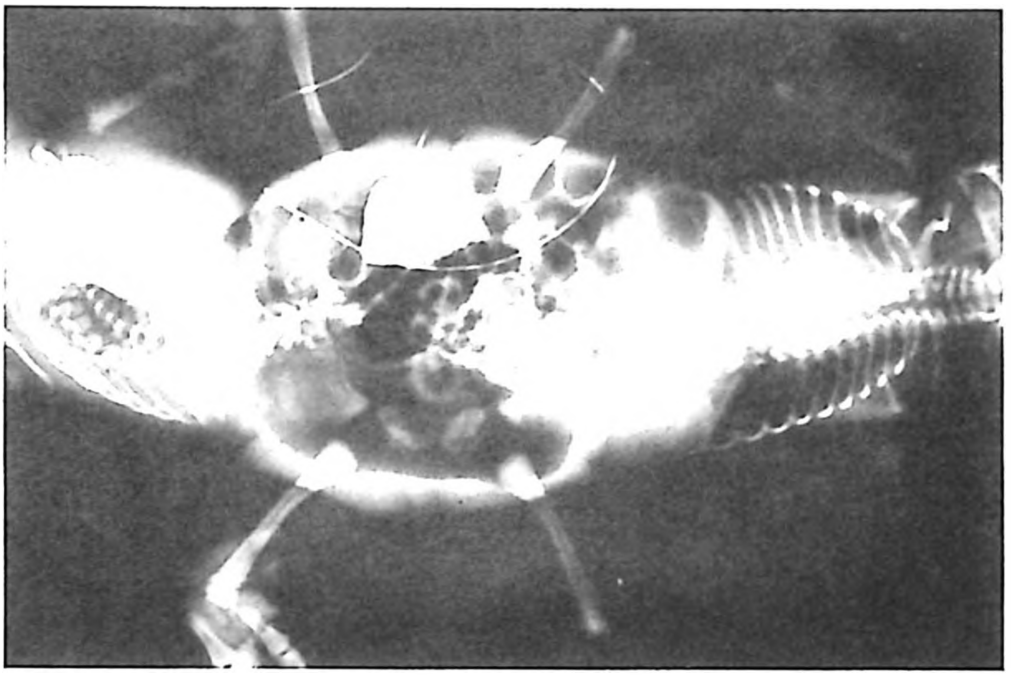

Figure 2

Intravenous pyelography of the twins.

the common pelvis. One ureter from each baby drained into the common bladders. In this way each bladder had a ureter opening into it from each of the twins (Figure 2).

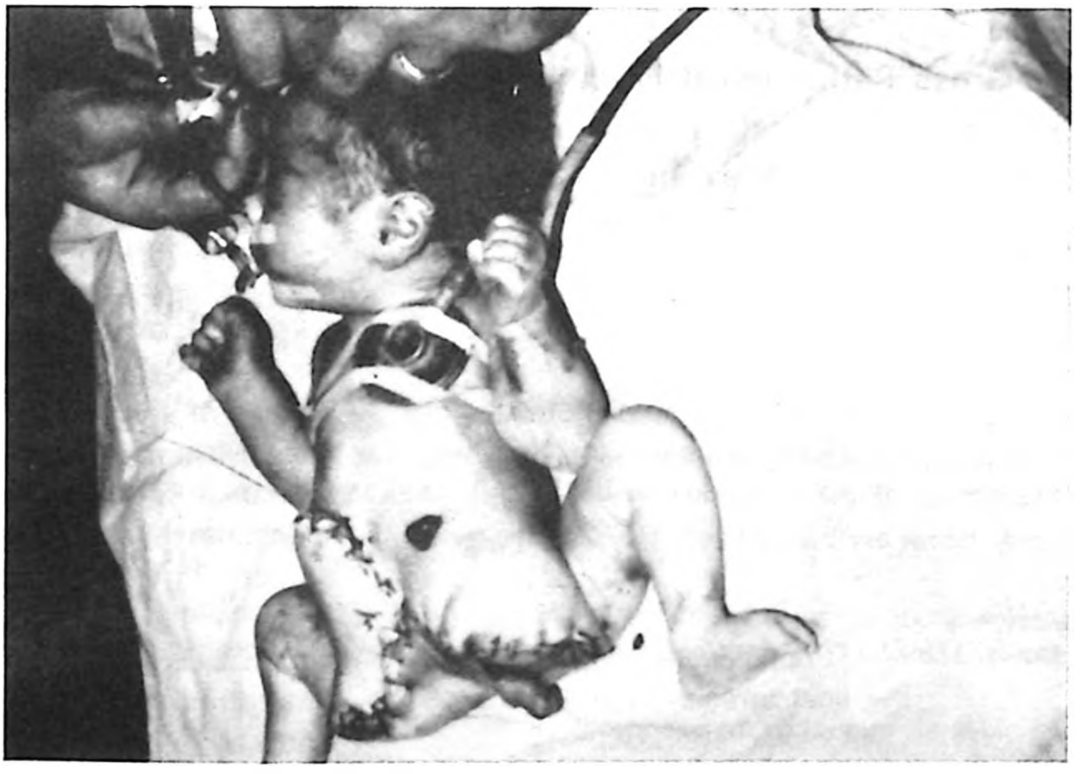

Figure 3

The early postoperative appearance of Baby I.

Since it was seen that Baby II's general condition was most unsatisfactory and affecting Baby I, it was decided to separate them under general anesthesia. Endotracheal intubation was unsatisfactory on Baby II, so the anesthetics were given to Baby I.

During the operation it was seen that at the level of the umbilical and iliac arteries the two babies has a common circulation, and on their separation, Baby II, whose general condition has been poor from the beginning, died. The exploration of the abdominal cavity showed the following pathological findings: ileal atresia, colonic agenesia, bilateral undescended hypoplastic testicles in Baby I; hypoplastic liver (one-third normal size) and hypoplastic spleen (half normal size), atresia of the anus, and bilateral undescended hypoplastic testicles in Baby II. IVP was also confirmed on exploration as well as the presence of two separate bladders one on either side of the common pelvis and the opening of the ureters into the opposite kidney (Table I). One of the bladders was taken out; a unilateral ureterocutaneostomy and an ileostomy were performed. The operation was completed by closing the abdcmen and the pelvis with flaps (Figure 3). Baby I died 14 hours after this surgical procedure.

On postmortem examination of each baby these further pathological findings were revealed (Table I): Baby I had an interatrial septal defect; Baby II had atresia of the larynx and trachea, bilateral lung hypoplasia, interatrial and high interventricular septal defects and hypertrophy of the right and left ventricles.

TABLE I

Gross Pathological Findings of the Ischiopagus Twins

	Baby I	Baby II
Head and Neck		– Pierre Robin abnormality – Hypoplasia of external ear – Atresia of larynx
Thorax	– Interatrial septal defect – Hemivertebra	– Atresia of trachea – Hypoplasia of lungs Interatrial septal defect – High interventicular septal defect
Abdomen	– Omphalocele – Ileal atresia – Colonic agenesia – Anomaly of urinary system – Bilateral hypoplastic undescended testicles	– Omphalocele (common) – Anal atresia – Anomaly of urinary system – Bilateral hypoplastic undescended testicles

Discussion

The types of conjoined twins most usually seen are thoracopagus, xiphopagus or omphalopagus, pygopagus, ischiopagus and craniopagus[1]. Conjoined twins are thought to be monozygotic, monochorionic twins of the same sex with identical chromosome patterns. It is believed that the malformation occurs soon after the formation of the rudimentary amniotic sac in the blastula stage. At this stage the embryonic dise divides into two separate and equal parts. If there is abnormal budding or fission of the embryonic disc, malformation occurs[1,3,5,9].

In cases where the condition of both twins is good, a delay in surgery in order to permit detailed examination is advantageous[1,7,10]. However, if one twin threatens the life of the other, early operation becomes mandatory[1,7]. In our case the second baby's general condition was very poor and was having an adverse effect on the first baby. For this reason the twins were taken into surgery immediately. During endotracheal intubation it appeared that Baby II had tracheal atresia and it was suspected that he also had hypoplastic lungs, so all efforts were directed toward saving Baby I.

This was the first time in Turkey that conjoined twins have been operated on, and it is sad that neither baby survived. Baby II with his serious congenital malformations, had no chance of living and Baby I would only have had a chance of survival if the operation had been performed before the negative effects of the other baby had begun to affect him.

Summary

A case of conjoined ischiopagus twins is described. This is the first case surgically separated in Turkey. Although it is advisable to delay the operation to allow time for diagnostic testing to determine the degree of fusion and the extent of malformations present, we had to operate without delay in view of the negative effects of the one on the other.

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