The Use of Radionuclides in the Investigation of Conjoined Twins

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Three sets of thoraco-omphalopagous conjoined twins were investigated. The clinical findings indicated a complex shared heart in two sets, and separate cardiovascular systems in the other. These assessments were confirmed by dynamic imaging after bolus injection of Tc-99m(Sn)colloid or Tc-99m millimicrospheres. Static images were then used to establish the configurations of the shared livers. The results of these studies were in keeping with the angiographic and autopsy findings in the two sets with complex cardiac anomalies and with the surgical findings during successful separation of the third set.

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About 75% of conjoined twins are thoraco-omphalopagous. Of these, conjunction involves the gastrointestinal tract in 50%, the pericardium in 90%, the heart in 75%, and the liver in 100% (*1*,*2*). The feasibility of surgical separation and the subsequent survival of the separated twins depends largely on the extent to which organs are shared, the nature of any other congenital anomalies, and acquired conditions such as birth trauma and infection.

Radionuclides have been used in the preoperative assessment of several sets of twins. The investigations include liver scanning with I-131 microaggregates of albumin (*3*), Tc-99m sulphur colloid (*2*,*4*,*5*) rose bengal (*6*,*7*), and Tc-99m dimenthyl-HIDA (*8*). The extent of crossed circulation has been measured by inhalation of $^{15}O_2$ (*9*) and by injection of labeled albumin (*10*,*11*) or erythrocytes (*4*,*11*,*12*). Technetium-99m has been used for dynamic imaging of the heart followed by delayed images of the liver and kidneys (*5*). Renal imaging with Tc-99m DTPA was performed in a set of twins who developed kidney failure (*5*).

In most of these reports, organ imaging and measurements of crossed circulation have been treated as separate investigations. This probably stemmed from limitations imposed by the available instruments and radiotracers. Petersen and Hill pointed out that quantitative volume-exchange measurements are of limited value since they provide no information on the nature of the vascular communication (*10*).

We conjectured that a colloid suitable for liver scanning would be the agent of choice in the initial assessment of thoraco-omphalopagous twins. It would establish the configuration of the liver and provide an indication of the magnitude and nature of any cross-circulation. After injection of colloid into one twin, radioactivity in the other would indicate the magnitude of the shunt, while the distribution of activity in the circulations and livers of the twins could provide information of the nature of the shunt. Activity in Twin 2 restricted to that part of liver contiguous with Twin 1 would be expected if crossed circulation was limited to the sinusoidal level. Uniform uptake in Twin 2's liver without significant systemic activity would be in keeping with portal-vein communication. Systemic and/or pulmonary activity would be an indication of a cardiac, arterial or venous shunt (other than portal).

This paper describes the investigations performed on three sets of thoraco-omphalopagous twins. The results were confirmed at autopsy in two sets and during successful surgical separation in the other. A review of conjoined twins admitted to our hospital between 1957

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and 1982 includes short case reports (Nos. 4,5, and 6) of these twins (*13*).

METHOD

The twins were placed in the lateral position under the detector of a gamma camera (30-cm field of view) fitted with standard 140-keV collimator. Radiocolloid was injected i.v. at the dorsum of one hand in Twin 1: Tc-99m albumin millimicrospheres or Tc-99m(Sn)colloid, 40.5 μCi (1.5 MBq) per kg of combined weight. Data were displayed on a television monitor and recorded for 4 min on floppy disc in a 64 × 64 matrix at 2 sec/frame. The available data processor determined the size of the matrix and the number of frames that could be stored on a disc. The frame rate was a compromise between these factors, the expected count rate, and the likely circulation time.

About 10 min after the injection of the colloid, posterior and lateral images of the liver were recorded with both twins in the field of view. Vertex views were recorded in the second and third sets of twins.

CASE REPORT 1

Clinical data. These twins were studied at 6 days of age. They had a combined weight of 4 kg and were joined from the midsternum to a common umbilicus. Each twin appeared to have one umbilical vein and two arteries. Respiration, suckling, and micturition were asynchronous. Clinical assessment and electrocardiogram (ECG) indicated a complex shared heart; both twins were cyanosed in 40% oxygen. Radiographs showed a shared lower sternum and (with barium) separate G.I. tracts down to the lower duodenum, where there was a communication.

RESULTS

Tracer reached both anterior mediastina simultaneously (Fig. 1), then promptly entered both systemic circulations. Twin 1 showed more circulating tracer than Twin 2, particularly in the pulmonary circulation. Paired regions of interest (ROIs) in the anterior mediastina, heads, and abdomens below the liver all showed twice as much activity in Twin 1. During the first 10 sec after appearance of "anterior mediastinal" activity, tracer elsewhere in the thorax of Twin 2 increased from <1% to 20% of that in Twin 1.

The static lateral and posterior images showed a globular liver, lying mainly within Twin 1. Both spleens were visible.

Colloid was then injected as a bolus into a vein in Twin 2's left foot, and the procedure repeated. In Twin 2 the inferior vena cava lay well posterior to the line of conjunction. Activity reached Twin 2 heart and Twin 1 anterior mediastinum simultaneously. Activities in anterior mediastinum, head, and abdomen of Twin 2 were now twice those in Twin 1—the converse of the earlier test. Densities in the liver and the posterior thorax, however, were not influenced by the route of injection.

Additional nuclear medicine procedures. On the 8th day of life Tc-99m *p*-butyl-IDA was injected into Twin 1. Excretion was slow, but at 45 min it showed separate

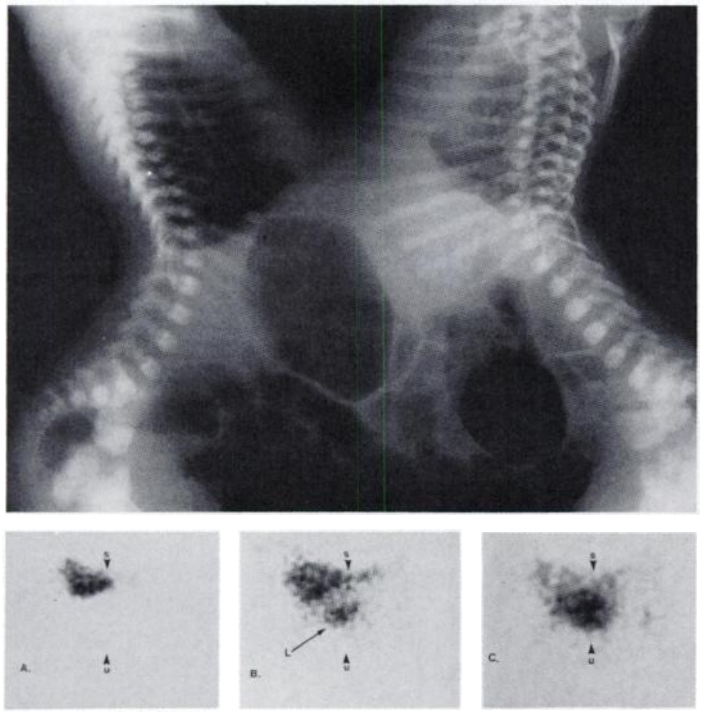

FIG. 1. Case 1. Right lateral images of Twin 1 recorded (A) 4 sec, (B) 6, and (C) 10 sec after injection of colloid into Twin 1. Arrows labeled s and u mark sites of conjunction at sternum and umbilicus respectively. Arrow L points to liver. Twin 1 is at left and Twin 2 at right. Radiograph was taken in same position.

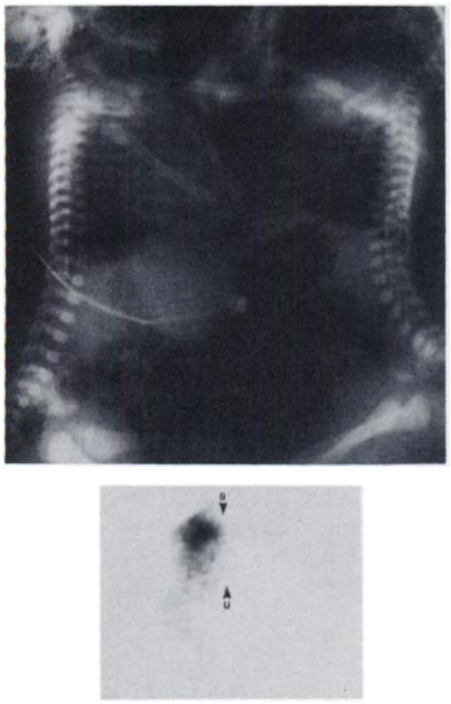

FIG. 2. Case 2. Right lateral image of Twin 1 formed by adding first five frames after entry of tracer into heart. Arrows labeled s and u mark the sites of conjunction at sternum and umbilicus. Twin 2 (not visible) lies to right of arrows. Radiograph was taken with a similar view.

biliary systems down to the gut. Posterior and lateral views at 90 min, suitably processed, showed about normal right/left lobe sizes in the liver of Twin 1, but Twin 2 had a very small left lobe.

Interpretation. The results suggested major shunts between the twins at atrial or pulmonary artery level, right-to-left shunts in each twin, a markedly reduced or absent pulmonary circulation in Twin 2, a common liver, and separate common bile ducts.

Cardiac catheterization. Cardiac catheterization on day 10 demonstrated a common atrium and probably a common sinus venosus, with all venous drainage to this chamber. Each twin had one atrioventricular valve and one ventricle. Twin 1 had a double-outlet ventricle with transposed great arteries, and Twin 2 had pulmonary atresia with a patent ductus arteriosus.

Autopsy. The twins died on the 54th day. At necropsy Twin 2 had a hypoplastic left lung.

Heart. The twins had separate aortae and venae cavae but a fused heart. Separate right and left superior venae cavae of each twin drained into a common "right" atrium; this communicated with a common "left" atrium that drained the two sets of pulmonary veins, and the two inferior venae cavae drained into the bridging area. Each twin had a single ventricle, receiving blood from its atria by separate valves. The ventricle of Twin 1 had a double outlet: a normal aorta but pulmonary hypoplasia. The ductus was patent. The ventricle of Twin 2 had a single aortic outlet with normal aorta. The pulmonary valve was atretic, and the ductus arteriosus was patent.

Liver. There was fusion between the left lobe of Twin 1 and the right lobe of Twin 2, with the division displaced towards Twin 2. The separate gallbladders led to separate cystic ducts and bile ducts. Although the pancreases were separate, the two common bile ducts joined before entering the single ampulla of Vater just below the duodenal fusion.

CASE REPORT 2

Clinical data. The twins, with combined weight 4.27 kg, were joined from the midsternum to a common umbilicus. Each had an umbilical vein and two arteries. Clinical assessment and ECG indicated separate ventricular and arterial systems. Barium meal and follow-through showed separate gastrointestinal tracts. The twins were investigated on the 9th and 10th days of life.

Results. With tracer in Twin 1, activity appeared in the pulmonary circulation, then in the systemic, with no evidence of any crossed circulation between the twins (Fig. 2). The static views of Twin 1's liver showed normal left and right lobes with only a small portion crossing the line of conjunction (Fig. 3).

Next day Twin 2 received the tracer in a repeat procedure. The absence of significant crossed circulation was confirmed; the pulmonary circulation filled before the systemic. Image processing (normalization, subtraction, and overlay) indicated that the inferior aspect of Twin 2's right lobe was slightly flattened and was fused with Twin 1's liver at the junction of left and right lobes (Figs. 4 and 5).

Interpretation. The impression was of separate cardiovascular systems, with no evidence of right-to-left shunt in either twin. There was a relatively minor crossed circulation between the two livers, probably at sinusoidal

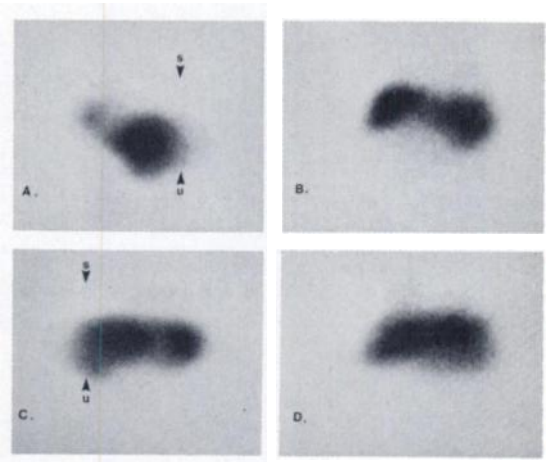

FIG. 3. Case 2. (A) right lateral, (B) posterior (C) left lateral, and (D) anterior views of Twin 1's liver. Arrows labeled s and u mark sites of conjunction at sternum and umbilicus.

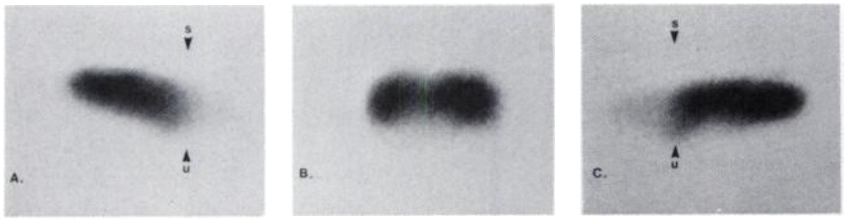

FIG. 4. Case 2. (A) right lateral, (B) posterior, and (C) left lateral views of Twin 2's liver. Arrows labeled s and u mark sites of conjunction at sternum and umbilicus.

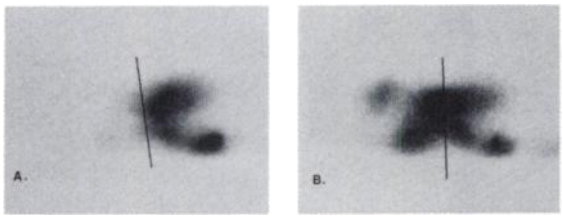

FIG. 5. Case 2. Vertex views of (A) Twin 2's liver, and (B) livers of both twins. Line is drawn through plane of conjunction.

level. Although the blood supplies were essentially separate, the livers were contiguous and/or fused over a large area.

Operative findings. These twins were successfully separated when they were 37 days old. The pericardium was shared but there was no major vascular communication. The livers were joined through the falciform ligaments. The peritoneal cavities were separate.

CASE REPORT 3

The combined weight was 5.67 kg. These twins were joined from the manubrium to a common umbilicus. Each had two umbilical arteries and one vein. Respiration was paradoxical. Clinical assessment and ECG indicated a shared heart. Lateral radiographs showed a horizontal shared manubrium. Barium meal and follow-through demonstrated separate gastrointestinal tracts. The twins were investigated on day 4.

Results. There was immediate total mixing, pulmonary and systemic, as soon as tracer reached the right side of the heart (Fig. 6). Time-activity curves of ROIs for the thorax and the abdomen below the liver were identical for the first minute after injection. The static views showed an extensive connection involving both lobes of the liver (Fig. 7).

Interpretation. The results suggested free communication between the pulmonary and systemic circulations of each twin, probably at atrial and/or ventricular levels. A repeat run, with Twin 2 receiving tracer, was not indicated.

Cardiac catheterization. There was free atrial and ventricular communication between the twins. The common atrium supplied two double-outlet single ventricles of the left-ventricular type in Twin 1 and right-ventricular in Twin 2. The ventricles communicated through a ventricular septal defect.

Autopsy. The twins died on day 75. There was a shared

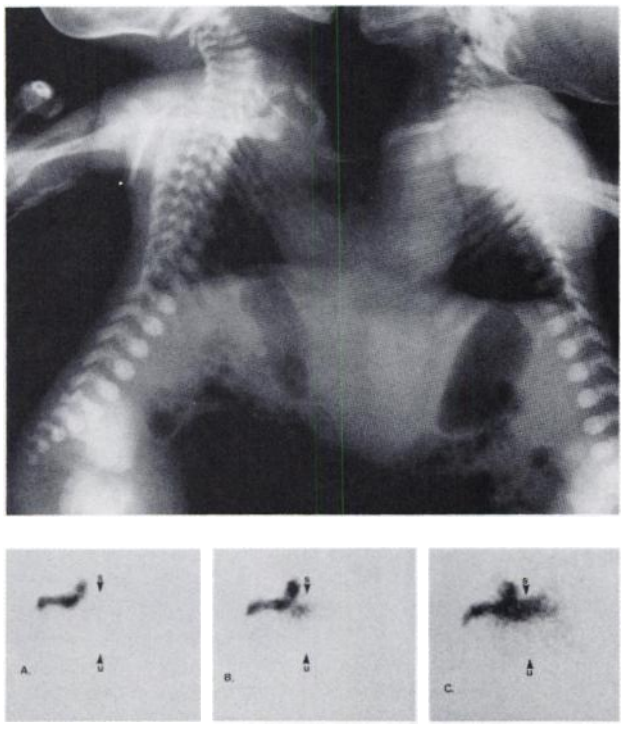

FIG. 6. Case 3. Right lateral images of Twin 1 recorded (A) 2, (B) 4, and (C) 6 seconds after injection of colloid into Twin 1. Arrows labeled s and u mark sites of conjunction at sternum and umbilicus. Twin 1 lies to left of arrow. Radiograph was taken in same position.

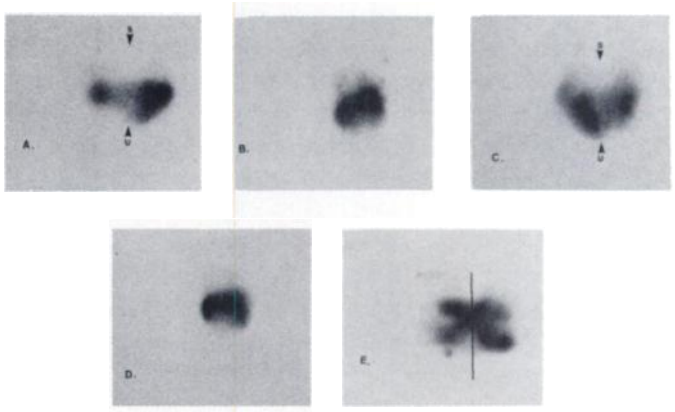

FIG. 7. Case 3. (A) right lateral, (B) posterior, and (C) vertex view. Arrows labeled s and u mark sites of conjunction at sternum and umbilicus. Line is drawn through plane of conjunction.

liver and a complex cardiac malformation. The left side of Twin 1's heart was fused to the right side of Twin 2's heart. Twin 1's pulmonary veins drained into a common venous mixing chamber, and superior and inferior venae cavae into the common atrium. Twin 2 had a single atrium that was in free communication with the venous mixing chamber. There was a patent foramen ovale between the twins' atria. Twin 1 had two ventricles linked by an atrioventricular canal defect that involved the mitral and tricuspid valves. Twin 2 also had two ventricles, the left hypoplastic; also a ventricular septal defect and mitral atresia. The left ventricles communicated through a ventricular septal defect.

DISCUSSION

The studies of these three sets of twins support the view that colloid is a suitable agent for the initial assessment of thoraco-omphalopagous twins. They also show that a more flexible data-storage facility would have made interpretation easier and might have made it possible to extract more information from the studies. In Cases 1 and 3 the immediate massive crossed circulation between the twins and the right-to-left shunts were obvious on the monitor; so was the absence of pulmonary or systemic cross-circulation and shunting in Case 2. The extensive sharing of the livers in Cases 1 and 3 was also readily apparent. But the analyses of the dynamic images in Case 1 and of the static images in Case 2 were much more difficult. As there was no clear demarcation between cardiac and pulmonary activity in Twin 1 of Case 1, the boundaries of the anterior mediastinal and posterior thoracic ROIs were arbitary. Demarcation might have been clearer if it had been possible to store the data in a larger matrix, in list mode, or at a higher frame rate. In Case 2 no single static image or series of images showed the plane of conjunction clearly, but it was fairly obvious after subtraction and overlay of selected images. It is possible that the exact location of the bridge would have been more obvious if a colloidal agent had been tagged, say, with Tc-99m for Twin 1 and with In-113m for Twin 2.

There is no doubt that radiotracer studies can play an important role in the evaluation of conjoined twins. The amount of vascular shunting and cross-circulation, or blood-volume exchange between the babies, can be established. Furthermore, organ-specific radiopharmaceuticals may be used to outline the degree of fusion of the liver. More specific details of the cardiac and vascular status can be obtained from angiographic studies.

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