Paediatrica Indonesiana 32 : 226 - 234 July - August 1992

CASE REPORT

Right Isomerism

by

ISMET N.OESMAN, HINDRA IRAWAN S.,
BAMBANG MADIYONO and SUDIGDO SASTROASMORO

(From the Department of Child Health, Faculty of Medicine, University of Indonesia)

Abstract

A rare case of complex heart malformatrion with right isomerism in a full-term baby boy which was the first case published in Indonesia is reported.

The patient was initially managed as a healthy baby, but on the 20th hours after delivery he repeatedly vomitted, and from the rontgenographic finding diagnosis at that time was duodenal atresia. During the duodenostomy operation the surgeon noticed that the spleen was absent. Cyanosis appeared after operation. Techocardiography and electrocardiography was done immediately, which showed complex cyanotic congenital heart disease. Diagnosis complex cyanotic congenital heart disease with right isomerism then was suspected. Unfortunately the baby died on the 12th day of admission. The cause of death was sepsis and anoxia because of complex cyanotic congenital heart disease. Autopsy findings confirmed the diagnosis of right isomerism (bilateral trilobes lung, bilateral right atrial appendages).

Received : Oktober 23, 1991

Introduction

It has long been recognized that there were cardiac anomalies associated with characteristically abnormal arrangements of the thoraco abdominal organs, particularly the spleen. On this basis, two distinct variants were recognized; one associated with absence of the spleen (asplenia sindrome) [1] and the other with multiple spleens (polysplenia syndrome) [2].

The fact that the organs in patients with asplenia had the basic arrangements of right isomerism, or bilateral right sideness', was first highlighted by Putschar and Mannion, and the left isomeric nature of patients with polysplenia was then emphasised by Moller et al [3].

Becker [4] stated that in the case of right atrial isomerism where absence of the spleen would be anticipated some of these patients had solitary spleens or even multiple spleens, while in cases with left atrial isomerism which would be anticipated to have multiple spleens, some of them had solitary spleen. Thus, the spleen is not a best guide to cardiac malformations, but atrial appendages which is most constantly present, is a better marker for detecting the cardiac lesion. He suggested not to use terminology the splenic syndromes but change it with "morphological method". Van Praagh et al [5] make a counter statement, that the concept of atrial isomerism in the heterotaxy syndromes with asplenia, or polysplenia or occasionally with normally formed spleen is anatomically erroneous. Because it has never been documented a case with morphologically right atrium appendages or left atrium appendages which were complete (not partial) mirror image of each other.

Here we describe a rare case of a male infant who demonstrated complex cyanotic congenital heart disease with right isomerism.

Case

A full-term baby boy was born spontaneously in our hospital without evidence of asphyxia. His birthweight was 2800 g and body length 50 cm. He was the fourth child of the family. His brother and sisters were healthy. The mother was referred to our hospital from a mid-wife because she suffered of polyhydramnions.

He was initially managed as a healthy baby, but he began repeatedly vomited on the 20th hours after delivery. Then a nasogastric tube was placed to aspirate the gastric fluid. Chest-X-ray and plain abdominal roentgenography were performed immediately. Fourty ml of gastric fluid were obtained and from the chest-X-ray we did not find esophagus obstruction, but abdominal roentgenogram showed double-bubble appearance. Diagnosis at that time was : doudenal atresia. Consultation to the Pediatric Surgery Department was done, and they planned to make an elective surgery on the third day of admission. To search any anomalies in another part of intestinal tract, a barium enema examination was performed, disclossing malrotation of the gut.

The surgeons made a doudenostomy, but during operation they found that the stomach and pancreas were right sided and the spleen was absent. After the operation the baby looked cyanotic; an ejection systolic murmur grade 3/6 on the left sternal border was heard. Electrocardiography was done and showed sinus rythm, left axis deviation and no signs of hypertrophy. From the echocardiography features there was complex

cyanotic congenital heart disease which consist of : transposition of the great arteries, complete AV canal and pulmonary stenosis. Further examination showed us that both inferior caval vein and the aorta patients ran on the right side of the spine and the right atrium-inferior vena cava junction could be visualized. At that time the diagnosis of right isomerism was suspected. On the seventh day we found Howell-Jolly bodies in the periphery blood, and on the eight day the baby looked clinically sepsis even the antibiotics had been given immediately after the operation. Blood culture we found Pseudomonas sp. On the twelfth day the baby passed away.

Autopsy was done and showed the spleen was absent, trilobus symmetrically lungs, transverse liver, stomach and pancreas were right sided. The heart was levocardia and showed complex heart malformation which consist of: triloculare biatrial, transposition of great arteries with patent ductus arteriosus and pulmonary stenosis, total AV canal and also anatomical bilateral right atrial appendages. So the diagnosis of right isomerism was confirmed.

Figure 1. *Complex cyanotic congenital heart disease (Trilokulare biatrial, complete AV canal transposition of the great arteries with patent ductus arteriosus and pulmonary stenosis also anatomic bilateral right atrial appendages*

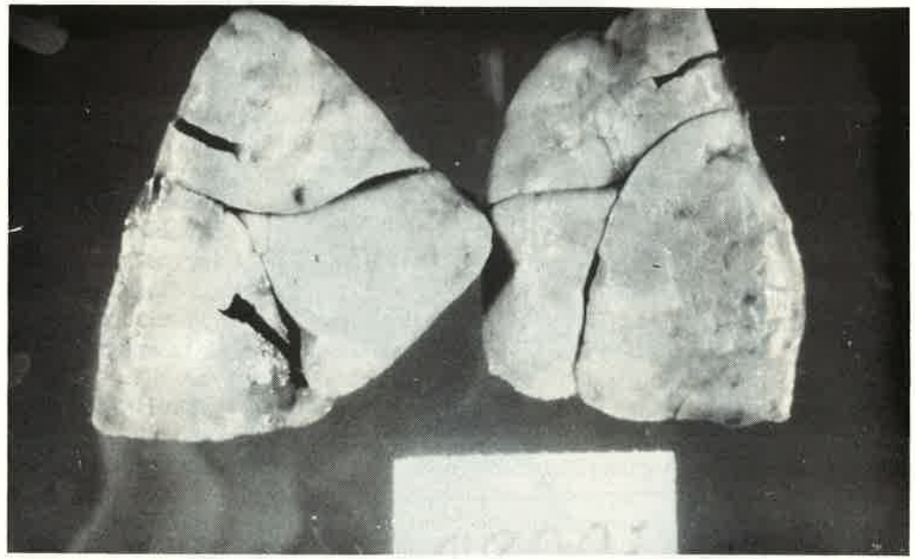

Figure 2. *Bilateral trilobed lungs*

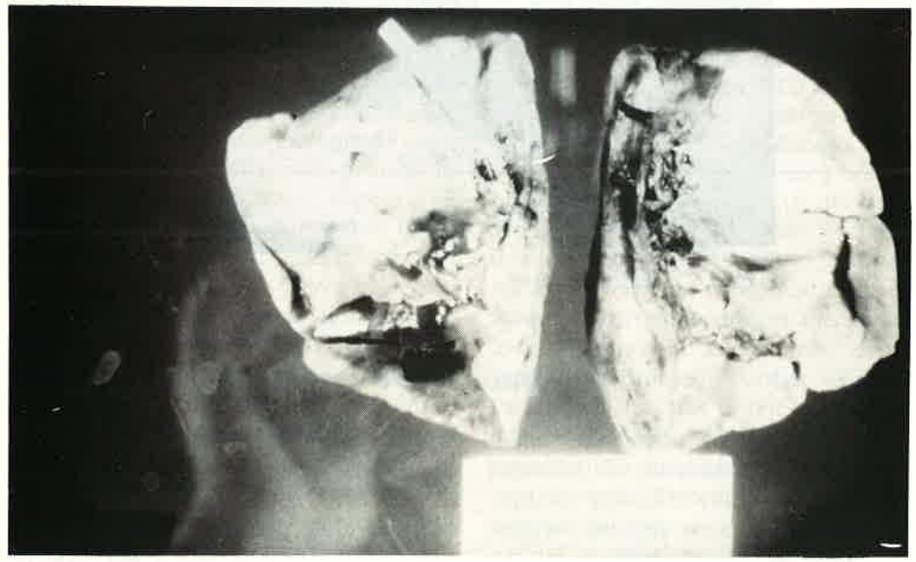

Figure 3. *Eparterial bronchi*

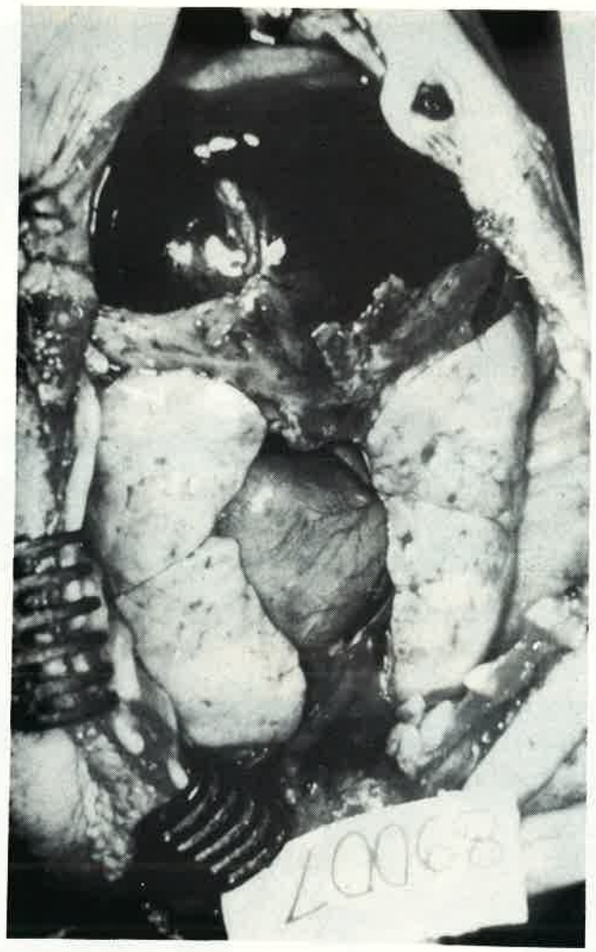

Figure 4. *Transverse liver*

Discussion

It has long been known that were combinations of malformations of the heart with characterisitically abnormal arrangements of the thoracoabdominal organs, particularly the spleen. Pohlius was the first man who described splenic agenesis in his patient in 1740 and Martin and Brechet in 1826 in separate reports describe the association of asplenia and cardiac anomalies [6].Since then, several cases have been described in various reviews, with IveMark in 1955 proposing an embryonic correlation of splenic agenesis and abnormalities of the heart and viscera [1]. This association with splenic abnormalities was considered so fundamental that it become conventional to name the cardiac anomalies on the basis of splenic morphology. Thus were born the syndromes of asplenia and polysplenia, or collectively the splenic syndromes.

Becker et al [4] stated that he found from 33 cases with left atrial isomerism, which would be anticipated to have multiple spleens, six patients had solitary spleens. In the case of right atrial isomerism, where absence of the spleen would be anticipated, nine patients of 72 had solitary spleens while one had multiple spleens. Thus, some of the cases in which splenic morphology was known, the arrangement of the spleen was discordant with the cardiac morphology. This investigation, therefore, showed a significant discord between splenic morphology and atrial arrangement in hearts having the characteristic cardiac malformations expected for the so-called "splenic syndromes'. So he suggest atrial appendages which is constantly present used as a determinant and describe abnormal arrangement of the heart and not to use again terminology splenic syndromes. But Van Praagh et al [5] in another report stated that the concept of atrial isomerism in heterotaxy syndromes is anatomically erroneous, beacuse of: first, a case with morphologically right atrium bilaterally, that is with bilateral inferior venae cavae, bilateral superior venae cavae, bilateral coronary sinus, bilateral septa secunda, and bilateral pyramid shaped appendages, has never been documented. Second, a case with morphologically left atrium bilaterally, that is with four pulmonary veins bilaterally, septum primum bilaterally, and finger-like atrial appendages bilaterally, has like wise never been documented. While in our case the atrium receives the inferior vena cava, the superior vena cava, and the ostium of the coronary sinus, it proved that our case has the morphologically right atrium.

Description of atrial isomerism simply means that there is duplication of those parts of the atrial chambers that exhibit the characteristic anatomical features of rightness or leftness. The heart with right atrial isomerism characterised by the presence of two atrial appendages each with morphology of the normal right appendage, and each having a broad and extensive junction with the smooth-walled components of the right atrial chambers as seen in our case [3].

It is not surprising, that the isomeric malformation should produce duplication of the atrial chambers without producing symmetry of the ventricles. Van Mierop et al [2], pointed out ,that the ventricles developed in series from the inlet and outlet parts of the ventricular 'loop', while the atrial chambers develop in parallel by incorporation of the venous sinus and primary pulmonary vein respectively into the right and left sides of the primitive atrial chamber.

It is difficult to calculate precisely the incidence of atrial isomerism because until recently it has been recognized most frequently at autopsy rather than during clinical examination. Sapire et al [3] were able to collect 45 cases from 2000 autopsy collections, and for our hospital this is the first case which published in Indonesia.

Visceral heterotaxy has long been recognised as the hall mark of the 'splenic syndromes'. The disposition of the abdominal organ led Ivemark to label asplenia as 'a syndrome of visceral symmetry'. Several subsequent authors then pointed to a midline liver as a marker of these 'splenic syndromes'. Further experience has shown that this is not the case, and that the abdominal organs are not usually arranged in symmetrical fashion. But our case showed bilateral trilobed lungs and symmetrical liver.

The morphology of the thoracic organs, specifically the bronchial tree, is a much better guide to the presence isomerism. Van Mierop et al [2] first observed that the bronchial arrangement was symmetrical in patients with isomerism. Bila-

14. Satomi G, Takao A. Systemic diagnostic method of two dimensional echocardiography in congenital heart disease. Heart and vessels 1985 ; 1 : 101-13.

15. Di-Donato R, Di-Carlo D, Squitieri C. et al. Palliation of cardiac malformations associated with right isomerism (asplenia syndrome) in infancy. Ann Thorac Surg 1987; 44: 5 - 39.