

COMPLETE TRANSPOSITION OF THE GREAT VESSELS

BY

C. J. INGOMAR AND E. TERSLEV

From the Dronning Louises Children's Hospital (Head: Professor Oluf Andersen), Copenhagen, Denmark

Received September 19, 1961

With the increased surgical possibilities of helping children with complete transposition of the aorta and pulmonary trunk the need for accurate diagnosis is increased, particularly regarding the site and size of shunts, because of their importance from the aspects both of operation and of prognosis. It is known that children with an accompanying patent ductus arteriosus in complete transposition have a shorter life span than those with a ventricular and/or atrial septal defect (Taussig, 1947; Hanlon and Blalock 1948; Astley and Parsons, 1952; Keith *et al.*, 1958).

In the individual patient neither the history nor the clinical picture are of particular significance in diagnosis. Radiological examination is of value because the heart often presents a characteristic appearance such as an "egg-shaped" silhouette with narrow vascular pedicle, and increased pulmonary vasculature (Fanconi, 1932; Taussig, 1947; Eek, 1949; Wood, 1950; Astley and Parsons, 1952; Keith *et al.*, 1953; Grosse-Brockhoff *et al.*, 1954; Kjellberg *et al.*, 1955; Miller *et al.*, 1958).

Cardiac catheterization does not usually provide conclusive evidence of the presence of complete transposition. Its importance may lie in the demonstration of veno-arterial communications (Campbell *et al.*, 1949; Blalock and Hanlon, 1950; Keith *et al.*, 1958).

Selective angiocardiology of the right heart undoubtedly is of the greater value in that it reveals the transposition (Goodwin *et al.*, 1949; Castellanos *et al.*, 1950; Cooley and Sloan, 1952; Astley and Parsons, 1952; Keith *et al.*, 1953; Grosse-Brockhoff *et al.*, 1954) and also helps to localize the shunt (Blalock and Hanlon, 1950; Gyllenswård and Lodin, 1954).

Venous angiocardiology is of no help in diagnosing shunts (Gyllenswård and Lodin, 1954). Its value is further diminished by failure to demonstrate the left side of the heart clearly (Goodwin *et al.*, 1949; Castellanos *et al.*, 1950; Keith *et al.*, 1953; Pung *et al.*, 1955).

The purpose of the present paper is to give an account of the diagnostic findings in 41 children in whom the diagnosis of complete transposition of the great vessels was made at the Dronning Louises Children's Hospital in Copenhagen between 1954 and 1960, and also to discuss diagnosis and differential diagnosis in relation to other heart diseases that are accompanied by permanent cyanosis.

RESULTS

One hundred and twenty-seven children with cyanotic congenital heart disease were admitted to hospital during this period. All were under the age of two years and so ill that it was considered vital to clarify the diagnosis. Thus, in addition to the routine clinical examination, all were investigated by cardiac catheterization and/or angiocardiology. The diagnosis and ages at the first examination are shown in Table 1.

The Diagnosis of Transposition. The patients with complete transposition comprised one-third, and this disease and the tetralogy of Fallot together comprised two-thirds, of the total series. Sixty of the 127 patients (who had the tetralogy or tricuspid atresia) were suitable for at least a palliative

TABLE I

127 CHILDREN WITH CYANOTIC CONGENITAL HEART DISEASE UNDER THE AGE OF TWO YEARS GROUPED ACCORDING TO THE FINAL DIAGNOSIS MADE BY THE DEPARTMENT

Department's final diagnosis	Total	Age at first examination		
		Less than 1 month	1-5 months	More than 6 months
Complete transposition	41	15	20	6
Fallot's tetralogy	44	9	14	21
Tricuspid atresia	16	2	7	7
Miscellaneous group	26	5	13	8
Total	127	31	54	42

operation, while in the miscellaneous group there were also six patients with pulmonary stenosis and atrial septal defect upon whom radical operations could have been performed. Thus there was a possibility of performing some form of surgical procedure in approximately half the 127 children studied under the age of two years.

Table I also shows that symptoms appeared very early in most of the children with transposition, so that diagnostic and surgical problems arose before the age of six months. The urgency of the problem was borne out by the fact that at the conclusion of the investigation 38 out of 41 patients had died, the average age of death being 4.9 months.

Eight of 17 children with complete transposition who came to necropsy did not have patency of the ductus arteriosus: their average age of death was 4.7 months, as compared with 1.8 months in nine children in whom the ductus was patent. Twenty of the 38 patients died before the age of three months and 30 before the age of six months.

In establishing the diagnosis it is important to use procedures that are as simple as possible, the aim being to arrive at a sufficiently accurate diagnosis by the usual clinical investigations, including routine radiological and electrocardiographic examinations. "Sufficiently accurate" is understood to mean that the diagnosis will permit a satisfactory decision regarding management of each patient in the light of the known course of the disease diagnosed.

TABLE II

ACCURACY OF THE CLINICAL DIAGNOSIS IN 127 CHILDREN UNDER TWO YEARS, WITH CYANOTIC CONGENITAL HEART DISEASE

Department's final diagnosis	Total	Number of correct clinical diagnoses
Complete transposition	41	22
Fallot's tetralogy	44	33
Tricuspid atresia	16	6
Miscellaneous group	26	1
Total	127	62

Table II shows the accuracy of the clinical diagnosis and indicates the number of patients in whom the cardiologist had made a diagnosis on clinical evaluation alone, and also the final diagnosis made as a result of angiocardiography and/or cardiac catheterization.

It was possible to make a correct clinical diagnosis in only 22 of the 41 patients with complete transposition of the great vessels. The tetralogy of Fallot was the only condition in this series in which it proved possible to make an accurate clinical diagnosis in the majority of children.

Thus it is clear that in most children under two years with cyanotic congenital heart disease who are so ill that operative intervention must be considered, the clinical diagnosis is unlikely to be sufficiently accurate to be relied upon alone.

A closer analysis of the findings upon which the clinical diagnosis was based is shown in Table III. Neither auscultation nor a history of cyanosis at birth or within the first week of life were of

TABLE III
CLINICAL AND RADIOLOGICAL FINDINGS IN 41 CHILDREN WITH COMPLETE TRANSPOSITION UNDER THE AGE OF TWO YEARS AT THE FIRST EXAMINATION

Clinical signs	No. patients	Per cent
Absence of cyanosis in the first week of life	15	37
Absent or soft murmur	20	49
Medium or loud murmur	21	51
Radiological signs		
Cardio-thoracic index less than 60 per cent	23	56
Narrow vascular pedicle	18	44
"Egg-shaped" cardiac silhouette	11	26
Pulmonary plethora	29	71
Narrow vascular pedicle and pulmonary plethora	18	44
Narrow vascular pedicle, "egg-shaped" cardiac silhouette, and pulmonary plethora	9	22
Concavity corresponding to the normal site of the pulmonary artery in the antero-posterior projection	10	24

value in the diagnosis of complete transposition. Radiological examination was of some value, for the combination of a narrow vascular pedicle, "egg-shaped" cardiac silhouette, and pulmonary plethora was found in 9 of 41 children with transposition but in only 2 of 86 children with other types of cyanotic congenital heart disease.

Cardiac catheterization and/or angiocardiography will be required in most patients to make a firm diagnosis. The value of catheterization in diagnosis might be questioned in the light of our experience, even when direct intubation of the anteriorly placed aorta arising from the right ventricle was possible. Angiocardiography with direct visualization of the aorta arising from the right ventricle, or of the pulmonary artery arising from the left ventricle (seen when contrast medium is injected into the left atrium) was of greater value. Seventeen of the patients who were diagnosed as having complete transposition, came to necropsy which confirmed the diagnosis in all.

The Diagnosis of Shunts. While transposition of the pulmonary trunk and aorta can be diagnosed clinically in approximately half the cases and by angiocardiography in the remainder, our results show that it was very much harder to detect the site of the shunt and to estimate its size. Clinical examination and radiological and electrocardiographic investigations offered little help in classifying the type of shunt (Table IV).

Since necropsy was performed in only two cases in which selective angiocardiography had been carried out, we cannot evaluate the value of this form of angiocardiography in the diagnosis of shunts. Table IV shows that venous angiocardiography was not sufficiently accurate in this regard, for it failed to demonstrate a shunt in 9 patients and proved correct in only 4 of 15 children so investigated.

In only one of nine patients proved to have a patent ductus was the patency diagnosed in life, which further emphasises the inadequacy of clinical investigations in diagnosing shunts. This inadequacy assumes importance in view of the unfavourable influence exerted by patency of the ductus upon prognosis.

It was concluded that neither venous angiocardiography nor cardiac catheterization were of significant value in diagnosing shunts. The prognostic implications of patency of the ductus are so

TABLE IV

THE DIAGNOSIS OF SHUNTS IN LIFE, THE NUMBER OF NECROPSIES PERFORMED, AND THE NUMBER OF CORRECT DIAGNOSIS CONFIRMED IN 41 CHILDREN WITH COMPLETE TRANSPOSITION UNDER TWO YEARS OF AGE

Diagnosis of shunts in life	Method of investigation (No. of necropsies in brackets)		Total no. of necropsies*	No. of correct diagnoses regarding shunts confirmed by necropsy
	Catheterization and angio- cardiography	Venous angio- cardiography only		
No shunt found	1 (0)	18 (9)	9	0
Shunts suggesting ASD or PFO	5 (1)	6 (4)	5	2†
Shunts suggesting VSD	2 (0)	3 (1)	1	1
Shunts suggesting PDA	0	1 (1)	1	1
Shunts suggesting combined ASD, VSD, PDA	5 (1)	0	1	0

ASD=Atrial septal defect. PFO=Patent foramen ovale. VSD=Ventricular septal defect. PDA=Patent ductus arteriosus.

* Two patients were examined by selective, and 15 by venous, angiography.

† Both patients were examined by venous angiography.

important that the use of catheterization or angiocardiology of the left heart may have to be considered. If the ductus is shown to be open, thoracotomy affords an opportunity of abolishing the shunt, which in the present state of our knowledge might be expected to prolong life provided there is a septal defect, until a more radical procedure could be performed.

SUMMARY AND CONCLUSIONS

A series of 41 children under two years of age with complete transposition of the great vessels, together with 86 children suffering from other forms of cyanotic congenital heart disease of the same age group, has been presented. The patients were investigated clinically, radiologically, and by cardiac catheterization and angiocardiology of the right heart.

Thirty-eight of the children with complete transposition died at an average age of 4.9 months. The importance of patency of the ductus arteriosus is emphasized by the earlier age of death, which averaged 1.8 months, in those with a patent ductus in addition to transposition.

Children with complete transposition did not differ clinically from other children with cyanotic congenital heart disease in the same age group, but radiological examination showed that the combination of a narrow vascular pedicle, "egg-shaped" cardiac silhouette, and pulmonary plethora in the antero-posterior projection was of value in diagnosis, and was present much more often in transposition than in other cyanotic lesions.

Angiocardiology was of considerable value in making the diagnosis but was of little help in analysing accompanying shunts.

REFERENCES

- Astley, R., and Parsons, C. (1952). *Brit. Heart J.*, **14**, 13.
 Blalock, A., and Hanlon, C. R. (1950). *Surg. Gynec. Obstet.*, **90**, 1.
 Campbell, J. A., Bing, R. J., Handelsman, J. C., Griswold, H. E., and Hammond, M. (1949). *Bull. Johns Hopk. Hosp.*, **84**, 269.
 Castellanos, A., Pereiras, R., and Garcia, O. (1950). *Amer. J. Roentgenol.*, **64**, 255.
 Cooley, R. N., and Sloan, R. D. (1952). *Radiology*, **58**, 481.
 Eek, S., in Mannheimer, E. (1949). *Morbus Cæruleus*. Basel. S. Karger.
 Fanconi, G. (1932). *Arch. Kinderhk.*, **95**, 202.

- Goodwin, J. F., Steiner, R., and Wayne, E. J. (1949). *Brit. Heart J.*, **11**, 279.
- Gyllenswärd, A., and Lodin, H. (1954). *Acta Radiol.*, **42**, 189.
- Grosse-Brockhoff, A., Schaede, A., and Lotzkes, H. (1954). *Deutsch. Arch. klin. Med.*, **201**, 305.
- Hanlon, C. R., and Blalock, A. (1948). *Ann. Surg.*, **127**, 385.
- Keith, J. D., Neill, C. A., Vlad, P., Rowe, R. D., and Shute, A. L. (1953). *Circulation*, **7**, 830.
- , Rowe, R. D., and Vlad, P. (1958). *Heart Disease in Infancy and Childhood*. New York, The Macmillan Comp.
- Kessler, A., and Adams, P. (1957). *Pediatrics*, **19**, 851.
- Kjellberg, S. R., Mannheimer, E., Rudhe, U., and Jonsson, B. (1955). *Diagnosis of Congenital Heart Disease*. Yearbook Publishers, Inc., Chicago.
- Miller, R. A., Baffes, T. G., and Wilkinson, A. A. (1958). *Pediatric Cardiology Index 1956/58*, 1109.
- Pung, S., Gottstein, W. K., and Hirsch, E. F. (1955). *Amer. J. Med.*, **18**, 155.
- Taussig, H. B. (1947). *Congenital Malformations of the Heart*. The Commonwealth Fund, New York.
- Wood, P. (1950). *Brit. med. J.*, **2**, 693.