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DIAGNOSIS *of* CONGENITAL HEART DISEASE

OF CONGENITAL HEART DISEASE

the Cardiologic Team of the Pediatric Clinic
Karolinska Sjukhuset, Stockholm

Published by

THE YEAR BOOK PUBLISHERS, INC.
CHICAGO

PRINTED IN U.S.A.

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DIAGNOSIS

A Clinical and Technical Study by

DIAGNOSIS OF CONGENITAL HEART DISEASE

Preface

THIS BOOK is based on clinical and roentgenologic studies of 396 cases of congenital heart disease investigated at the Pediatric Clinic of Karolinska Sjukhuset, Stockholm, between October 1951 and June 1954. They were carried out by a team consisting of two cardiologists and two roentgenologists, part of whose work at the clinic is the investigation of patients with symptoms and signs of cardiovascular disease.

An account is given of the results obtained with the diagnostic methods used, with particular emphasis on the special technical aids which make possible a detailed functional and anatomic diagnosis. Evaluation of the findings at the customary clinical examinations has been greatly facilitated by these means. The reason underlying the high incidence of extensive special investigations was to promote one of the main objects of this book, namely, to appraise the value of the respective methods.

The results of the physical examinations, including phonocardiography, have been analyzed by Edgar Mannheimer and Bengt Jonsson, and those of the other clinical investigations and of cardiac catheterization by Bengt Jonsson. Sven Roland Kjellberg and Ulf Rudhe are responsible for the interpretation of the roentgenologic and angiocardiographic findings, and Ulf Rudhe for the observations at electrokymography.

Space does not permit us to thank all those who have helped in the preparation of this book. We nevertheless wish to express our particular gratitude to the following:

To Arvid Wallgren, for his encouragement during the course of our work.

To Leo G. Rigler, Jack Friedman and Fred Jenner Hodges, who have shown great interest in our project.

To Clarence Crafoord, Gunnar Ekström, Viking Olof Björk and Åke Senning, for many stimulating discussions.

To Lennart Rahm, for valuable theoretical views on hemodynamics.

To Johan Bonnier, who with unfailing generosity has given us considerable financial assistance.

To Åke Wilton and Gunnar Ekström, who kindly placed autopsy specimens at our disposal.

To Erica Odelberg, who translated the book, as well as to the Year Book Publishers for their helpful co-operation.

—THE AUTHORS.

Stockholm
December, 1954

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1 Embryologic Survey

THIS CHAPTER has no pretensions to being a complete account of the whole course of development of the circulatory system. Its object is merely to elucidate, from the embryologic viewpoint, the malformations of the heart and great vessels that comprise the clinical material in our book. The data in this chapter are based mainly on current conceptions of the process of development (7, 54, 55, 64, 95, 112, 134, 290, 291, 313, 347). (See also the anatomic background in the relevant chapters.)

A. DEVELOPMENT OF THE GREAT VEINS

1. The Systemic Veins

The circulatory system is laid down at a very early stage of fetal development. According to Eternod (127), a closed blood circuit already exists in a human fetus about 1.3 mm long, at the end of the second embryonic week. This circuit consists of the heart, two aortae, which pass caudally into the umbilical arteries, and two *umbilical veins*. The task of these veins is to carry the blood from the capillaries of the chorion directly to the heart. Further development takes place rapidly, and in a subsequent stage branches grow out from the umbilical veins and unite with the newly formed vessels in the yolk sac (290, 291). These venous branches increase rapidly in size and form the so-called *omphalomesenteric* veins, which open into the umbilical veins slightly caudal to the heart. Because of the increased blood flow through the parts of the umbilical veins lying cranially to the site of entry of the omphalomesenteric veins, they become widened into a *sinus venosus* (Fig. 1).

The blood which circulates within the embryo is, on the contrary, collected by the so-called *cardinal veins*. These are laid down in the third embryonic week (128) as two paired venous trunks. One pair, the *anterior cardinal veins*, arises from the cranial part of the body; the other pair, the *posterior cardinal veins*, arises from the caudal part. These cardinal veins unite on either side of the heart into a short main trunk, the *common cardinal veins* or *ducts of Cuvier*. They, in turn, open into the previously mentioned sinus venosus (Fig. 2). Initially, the *posterior cardinal veins* collect the main part of the embryonic blood. This is because these veins drain not only the segmental veins of the thoracic, abdominal and pelvic regions, but also the veins from both the lower and upper extremities. As the heart descends caudally, the orifices of the *subclavian veins* are, however, displaced cranially, so that they will gradually open into the *anterior cardinal veins*.

After this occurrence, a wide anastomosis forms between the anterior cardinal veins: the *left innominate vein* (Fig. 2, *D–F*). The part of the right cardinal vein which lies caudal to the site of anastomosis

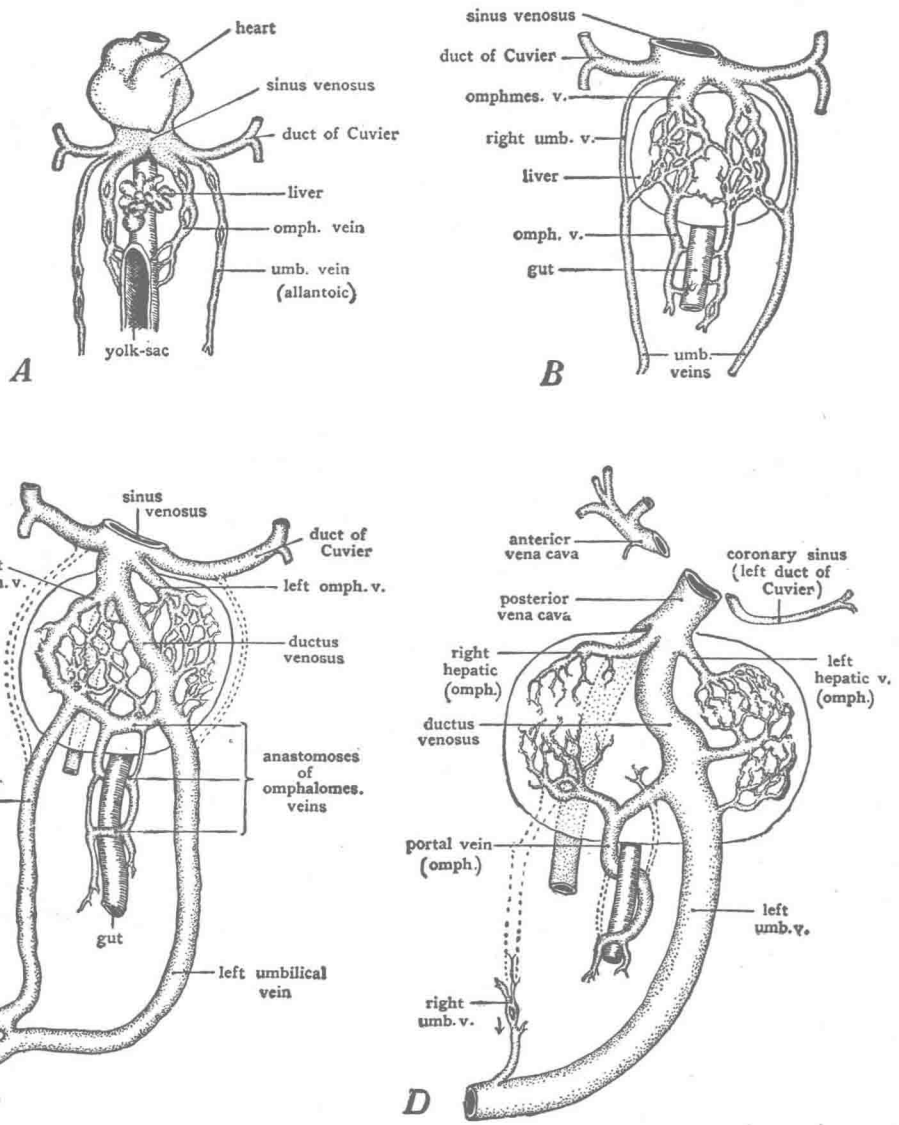

FIG. 1.—Diagrams showing development of hepatic portal circulation from omphalomesenteric veins, and changes by which blood returning from placenta by way of umbilical veins is rerouted through the liver. *A*, based on conditions in pig embryos of 3–4 mm, applicable to human embryos of fourth week; *B*, based on pig embryos of about 6 mm, applicable to human embryos of fifth week; *C*, based on pig embryos of 8–9 mm, applicable to human embryos in sixth week; *D*, based on pig embryos of 20 mm and above, applicable to human embryos of 7 weeks and older. (From Patten, B. M.. *Human Embryology* [New York: Blakiston Company, 1946].)

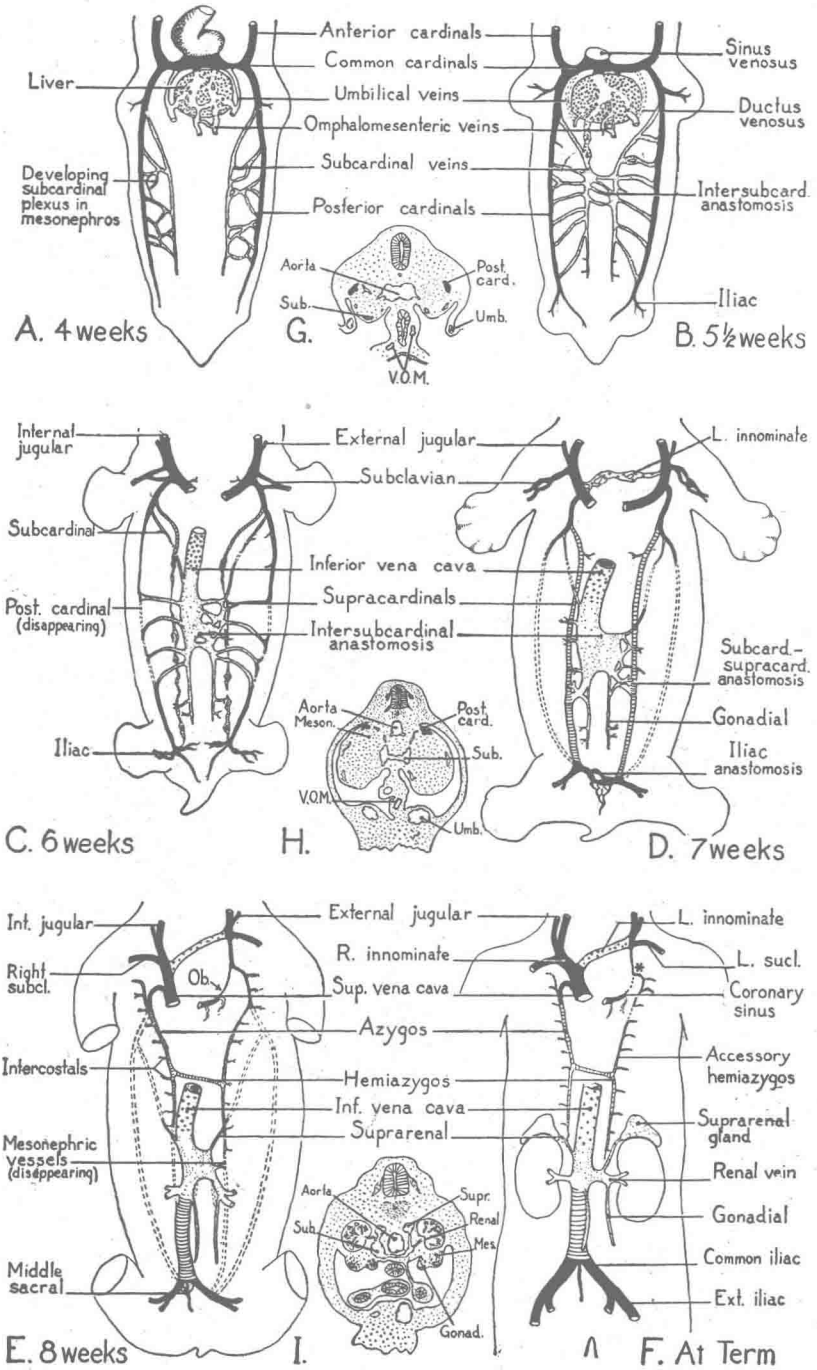

FIG. 2.—Schematic diagrams, ventral views, showing some of the steps in development of inferior vena cava. Cardinal veins are shown in black, subcardinal system is stippled, supracardinals are horizontally hatched; vessels arising independently of these three systems are indicated by small crosses. (From Patten, B. M.: *Human Embryology* [New York: Blakiston Company, 1946].)

forms, together with the common cardinal vein, the primordium of the *superior vena cava*. The part lying between the site of anastomosis and the right subclavian vein becomes the *right innominate vein*. At this stage, a *left superior vena cava* also exists. It is formed by the left cardinal vein, below the origin of the left innominate vein, and the left common cardinal vein. The greater part of this caval vein soon atrophies (137). Only the caudal part persists in the form of the *oblique vein of the left atrium* (Fig. 3), as well as the cranial part, in the form of the *first intercostal vein* (Fig. 4). We shall return to this matter on page 13.

Considerable deviations frequently occur. Sometimes no anastomosis takes place between the two anterior cardinal veins. The blood from the left upper part of the body then continues to flow downward through a *persisting left superior vena cava*, as shown in Figure 386 (p. 395). Even when an anastomosis is present, there may be *double venae cavae* (Fig. 5).

The fate of the *posterior cardinal veins* is so intimately associated with the development of the *hepatic veins* and the *inferior vena cava* that a short account of this process is necessary.

The two omphalomesenteric veins run through the mesenchymal septum transversum, which separates the caudal part of the pericardial cavity from the pleural and abdominal cavities (54, 250). In connection with the development of the medial lobe of the liver, two vascular networks are

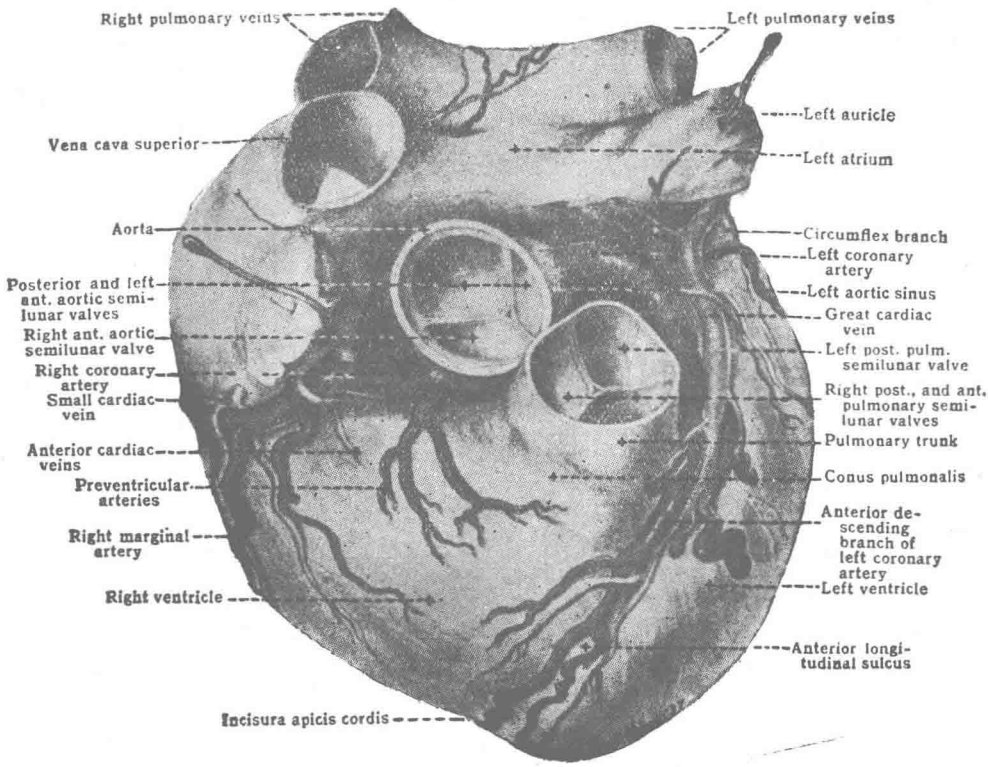

FIG. 3a.—*A*, cephalic view of the heart with the epicardium removed to expose the injected coronary vessels. (*A* and *B* from *Morris' Human Anatomy* [10th ed.; New York: Blakiston Company, 1942].)