The Salient Points and the Value of Venous Angiocardiography in the Diagnosis of the Cyanotic Types of Congenital Malformations of the Heart

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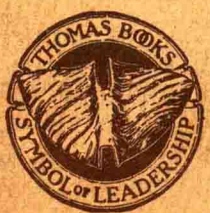

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PREFACE

This book is the result of a ten year study of 421 angiocardiograms, each one consisting of approximately twenty films, taken on 283 patients with cyanotic types of congenital malformations of the heart.* The salient points in the angiocardiographic diagnosis of ten entities are presented.

In order to best establish the value of angiocardiography as a separate laboratory tool, the authors based the angiocardiographic diagnosis of each entity only on the knowledge that the patients were cyanotic, and on the interpretation of the angiocardiographic findings.

The accuracy of this angiocardiographic diagnosis was then checked in each case against the final diagnosis arrived at by a complete physical, fluoroscopic, roentgenologic and electrocardiographic examination and, in addition, by cardiac catheterization and/or surgery and/or autopsy.

Twenty-one entities of the cyanotic type of congenital malformations of the heart are classified into four groups. We wish to emphasize that this manuscript represents only a summary of the basic findings of the most important types of congenital malformations of the heart.

The authors wish to extend their thanks and appreciation to Dr. Charles T. Dotter, Professor and Chairman, Department of Radiology of the University of Oregon Medical School, for reading the proofs and offering many helpful suggestions. We also wish to thank our chief technician, Mr. Glen Volz and our artist, Mrs. Gloria Jones who have faithfully assisted in this work since its inception.

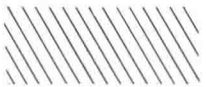
* The total number of angiocardiograms that the authors have studied on proven cases of various cyanotic and non-cyanotic types of congenital malformations of the heart is over 1,000.

CONTENTS

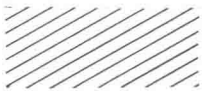
The Salient Points and The Value of Venous Angiocardiography in the Diagnosis of the Cyanotic Types of Congenital Malformations of the Heart

The following Key is for the drawings on pages 8, 17, 21, 28, 36, 44, 45, 51, 58, 61, 65.

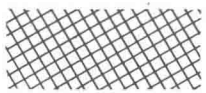
KEY



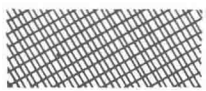
Venous blood



Arterial blood



Mixed blood predominantly arterial



Mixed blood predominantly venous

S.V.C.—Superior Vena Cava
I.V.C.—Inferior Vena Cava
R.A.—Right Atrium
R.V.—Right Ventricle
P.A.—Pulmonary Artery
P.V.—Pulmonary Veins
L.A.—Left Atrium
L.V.—Left Ventricle
A.O.—Aorta
C.T.—Common Trunk
Br.A.—Bronchial Artery

The figures in the cardiac chambers and in the vessels denote the respective percentage of their oxygen saturation.

INTRODUCTION

THE PURPOSE of this book is to bring out the salient points in the angiocardiographic diagnosis of the various cyanotic types of congenital malformations of the heart and to establish the value of angiocardiography as a diagnostic tool for these entities. We, therefore, based our diagnosis on angiocardiographic findings alone and the knowledge that the patients were cyanotic. In order to best establish the diagnostic value of angiocardiography as a separate laboratory tool, all other available information regarding the history, physical, fluoroscopic, roentgenologic, electrocardiographic, cardiac catheterization and autopsy findings was purposely omitted. During the past ten years 421 angiocardiograms, each consisting of an average of twenty films, were done on 283 patients with cyanotic types of congenital malformation of the heart. Ninety-five percent of our patients were under fifteen years of age. A complete physical, fluoroscopic, roentgenologic and electrocardiographic examination was done routinely on all patients before an angiocardiogram was performed. In addition to these examinations the diagnosis was confirmed in 149 patients by cardiac catherization, in 101 patients by surgery and in 98 patients by autopsy findings. As a result of this study, we believe that the diagnostic value of angiocardiography can be best demonstrated by classifying the cyanotic types of congenital malformation of the heart into four groups and by the presentation of their salient angiocardiographic points.

* From the Hektoen Institute for Medical Research, the Pediatric Cardiophysiology Department of Cook County Children's Hospital, the Pediatric and Surgical Departments of the University of Illinois College of Medicine and of the Presbyterian Hospital of Chicago, Illinois. This study was aided by a grant from the Chicago Heart Association.

Group 1

Entities in which the diagnosis can *ALMOST ALWAYS* be made by proper interpretation of a technically good angiocardiogram (Table 1).

Group 2

Entities in which the diagnosis can *USUALLY* be made by proper interpretation of a technically good angiocardiogram (Table 2).

Group 3

Entities in which the diagnosis can *USUALLY NOT BE MADE* by proper interpretation of a technically good angiocardiogram (Table 3).

Group 4

Entities which *ALWAYS REQUIRE* additional studies before a definite diagnosis can be established (Table 4).

The salient points in the angiocardiographic diagnosis of the following ten cyanotic types of congenital malformations of the heart which belong to our Groups 1, 2 and 3 will now be presented briefly. Eleven other types of cyanotic congenital malformations of the heart belonging to Group 4, which represent entities where the value of angiocardiography alone is limited, will not be discussed.

GROUP 1

TETRALOGY OF FALLOT

PATHOLOGY

1. Varying degrees of infundibular stenosis or combined infundibular and valvular stenosis. Rarely, only valvular stenosis which is then usually associated with post-stenotic dilatation of the pulmonary artery. Exceptionally, valvular or pulmonary atresia.

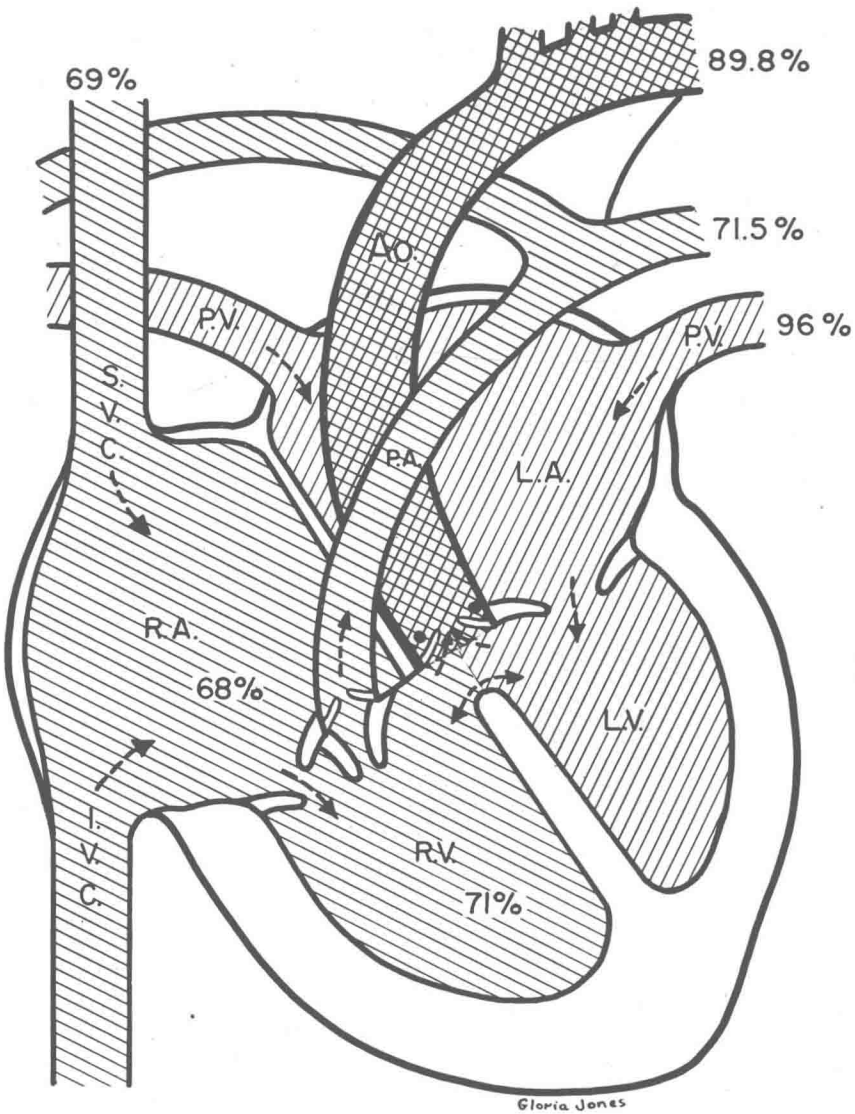
2. Large defect in the membranous and upper muscular portion of the ventricular septum.

3. Varying degrees of functional dextroposition (overriding of aorta.

4. Because of the above anomalies, the right ventricle is hypertrophied and, in some cases, dilated.

HEMODYNAMICS

Systemic venous blood returning to the right atrium and ventricle is diverted from the lung in proportion to the degree of obstruction caused by the stenosis. The venous blood which cannot enter the stenotic outflow tract will follow a path of lesser resistance and pass into the aorta. This produces cyanosis. The systolic pressure in the right ventricle is approximately the same as in the left ventricle. In all cases the pulmonary blood flow is decreased and the systemic flow is increased. In cases where the pulmonary artery is atretic, the pulmonary circulation is maintained by bronchial arteries or other collateral vessels.



TETRALOGY OF FALLOT

(See Key on page 2.)

Salient Angiographic Features

1. Simultaneous visualization of the aorta and pulmonary arteries (Fig. 1-a).

2. In the majority of cases the main pulmonary artery and the branches are hypoplastic and the aorta is wide. In about 20% of these cases there is a right aortic arch (Fig. 1-b).

3. Patients with valvular or combined stenosis usually reveal a post-stenotic dilatation of the pulmonary artery.

4. In some patients with localized infundibular stenosis associated with valvular stenosis, there is opacification of a third ventricle. The size of this third ventricle varies depending upon the space between the localized infundibular stenosis and the pulmonary valves (Fig. 1-c).

5. Opacification of the peripheral pulmonary vessels is diminished.

6. Faint, or no opacification of the left cardiac chambers. Usually the fainter the opacification the more severe is the malformation. The opacification of the collateral vessels in these severe malformations is oftentimes well visualized (Fig. 1-d).

7. The type of stenosis (valvular, infundibular or combined) could at times not be determined in our series (Fig. 1-e and 1-f).

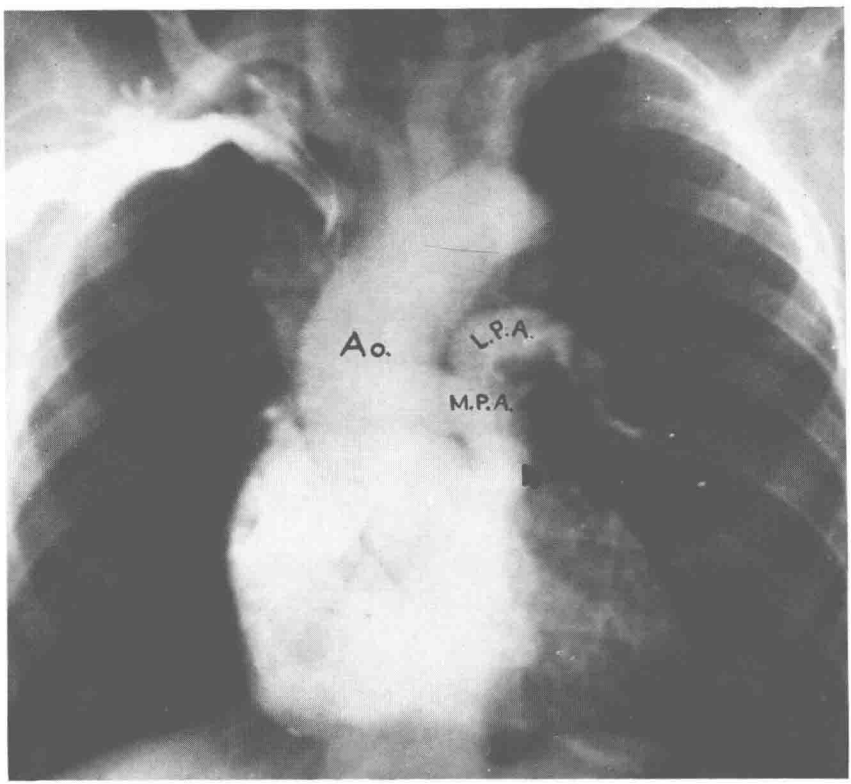


Figure 1-a. Tetralogy of Fallot. A-P view of a typical severe tetralogy of Fallot at 1½ seconds. Early and simultaneous visualization of hypoplastic pulmonary arteries and of a markedly dextroposed wide aorta. Note the location of the pulmonary semilunar valves (arrow); the opacification of the peripheral pulmonary vessels is diminished. MPA—main pulmonary artery; LPA—left pulmonary artery; AO—aorta.