INTERNATIONAL ACADEMY OF PATHOLOGY MONOGRAPH

THE HEART

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by 18 authors

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FOREWORD

This monograph, *The Heart*, is the 15th in the series of *Monographs in Pathology* initiated by the International Academy of Pathology in 1959. Each monograph represents a compilation of expanded lectures and illustrative materials born of the Academy's Annual Long Course. By assembling materials of the finest educational standards and values relative to normal and abnormal structure and function of organs and systems, the long courses and their offspring the monographs contribute measurably to the realization of the educational goals of the Academy. "The Pathophysiology and Anatomy of the Heart," which comprised the Long Course at the 62nd Annual Meeting of the United States-Canadian Division of the Academy in Washington in 1973, fully meets the goals and the expectations of the Academy. This monograph, a sequel and permanent record of that course, encompasses not only the lectures presented at it, but substantial additional material.

The Directors of the Long Course, Dr. Jesse E. Edwards and Dr. Maurice Lev, assembled an impressive faculty of scientists and clinicans and their presentations in this monograph form a rich deposit of knowledge for the use of all who have interest in or seek further understanding of the cardiac diseases. The vast amount of investigative work, both basic and clinical, that has led to this present distillate is awesome. Each chapter contributes immensely to our present day understanding of heart disease and is an individual monument to the authors' investigations and industry.

The Academy expresses its sincere appreciation to Dr. Edwards and Dr. Lev and to their distinguished faculty who contributed their knowledge and effort to this monograph, and to the publishers, The Williams & Wilkins Company, for their support.

MURRAY R. ABELL, M.D., PH.D.
Series Editor

PREFACE

The fields of congenital and acquired cardiac disease are ever widening. More accurate definitions of standard conditions have characterized pathologic studies as dictated by refinement of clinical diagnoses and introduction of operative procedures. Progressive sophistication in the field of Cardiology has served to make the pathologist more accurate in his distinction of one condition from another.

This symposium attempts to bring up to date the knowledge that has been acquired in recent years as addition to or correction of standard descriptions of the past.

Beginning with fundamentals on development of the heart, its function and hypertrophic responses, the symposium then concerns itself with a variety of specific subjects dealing with acquired diseases of the circulatory system and cardiovascular malformations.

As a handbook, the material will have value to the pathologist in his day-to-day work. Beyond this, as pathology is, in essence, part of clinical medicine, it is hoped that the material presented will have meaning as a foundation for those dealing with the diagnostic and therapeutic sides of Cardiology.

The bibliographies found in the individual chapters should be a suitable guide to the student of the subject who cares to probe more deeply into any of the subjects presented.

The editors take pride in the fact that each of the authors is an expert in his field, a state reached by long time involvement which continues to the present.

We are indebted to each of these authors for having given of their time, energy and knowledge without which this symposium would not have been possible.

Our gratitude goes also to the International Academy of Pathology for having provided the fertile soil upon which the seeds of knowledge could be planted.

JESSE E. EDWARDS, M.D.
MAURICE LEV, M.D.

Contents

Chapter 1

Anatomy and Embryology of the Right Ventricle

L. H. S. VAN MIEROP

Of the four cardiac chambers, the right ventricle is anatomically, phylogenetically, and developmentally the most complex. This undoubtedly is related to the development in phylogeny of pulmonary respiration with the establishment of two parallel circulations.

From the clinician's point of view, the right ventricle is also the most important chamber, since most congenital cardiac anomalies, particularly the more complex types, involve the right ventricle in some way. In the great majority of cases it is the right ventricle which is entered in carrying out surgical procedures aimed at the functional correction of anomalies involving this chamber or the ventricular septum, or both.

ANATOMY

The term "right" ventricle only really applies if the heart is removed from the body and placed on its apex with the cardiac septum in a sagittal plane. With the heart lying in its natural position in the intact body, the right ventricle is actually located anteriorly, or at best, anterior and slightly to the right. Similarly, the posterior (diaphragmatic) wall of the heart is in reality located inferiorly.

The normal right ventricle is roughly pyramidal in shape (Fig. 1). The anterior or sternocostal wall is somewhat convex, the left posterior or septal wall concave and the inferior or diaphragmatic (usually referred to as "posterior") wall is nearly flat. The right posterior "wall" is formed by the plane of the atrioventricular ostium and the crista supraventricularis. The ventricular walls as such remain unnamed in both the *Basle Nomina Anatomica* (*B.N.A.* 1895) and the third edition of the *Nomina Anatomica* (*N.A.* 1966).

The right ventricular cavity (Fig. 2) is traditionally divided arbitrarily into a posteroinferior inflow portion containing the tricuspid valve apparatus, and an anterosuperior outflow portion from which the pulmonary trunk arises. Inflow and outflow portions are separated from each other by a complex of more or less well defined muscular bands which together form a not quite complete circular orifice. Some disagreement still remains at present concerning the nomenclature of its component parts. The most prominent and constant part is represented by

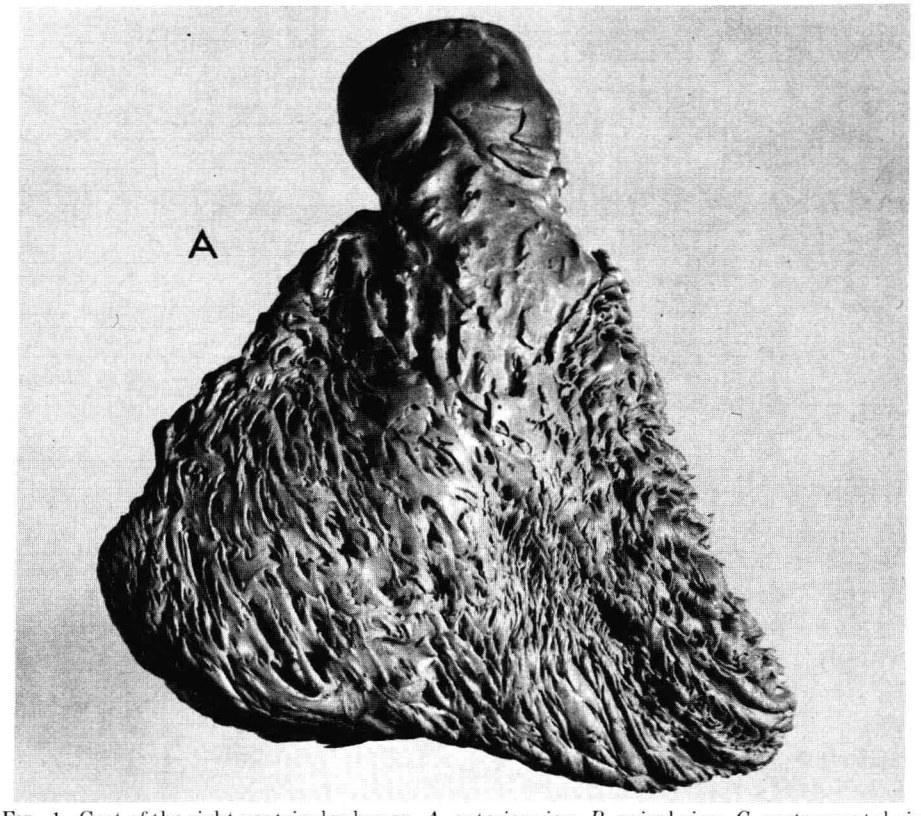

FIG. 1. Cast of the right ventricular lumen. *A*, anterior view. *B*, apical view. *C*, posteroseptal view. *RAVO*, right atrioventricular ostium. *PR*, root of pulmonary trunk. *CSV*, concavity caused by the crista supraventricularis.

a well defined, saddle-shaped muscular band which separates the tricuspid and the pulmonary valves. In both the *B.N.A.* and the *N.A.* this structure, first described but not named by Wolff in 1781,[19] is called the crista supraventricularis. While the name "crista supraventricularis," according to Mall[10] introduced by His in 1886,[3] has long been generally accepted, there still is some confusion as to its application. Keith, in 1909,[4] did not mention it, but described and illustrated a right and a left infundibular band which join each other below (proximal to) the pulmonary valve (Fig. 3). The line of junction, according to Keith, may be visible in some normal and abnormal hearts as a shallow groove, called a raphe by Keith. The right band is described as running to the right towards the base of the right ventricle, the left band toward the apex. Keith further states that "the interbulbar septum (the bulbar or conus septum of other authors) is laid down at first as an endocardial fold into which the two muscular bands . . . push their origins." He does not, therefore, consider the infundibular bands themselves to be actual derivatives of the embryonic bulbus or conus cushions, as some authors have suggested. A shallow, weakly S-shaped groove running from the commissure between the posterior pulmonary cusps or base of

the left cusp and the base of the medial papillary muscle is indeed present in many normal and abnormal hearts, particularly in those of infants. It is thought by some to represent the line of fusion of the embryonic conus cushions, which would be expected to terminate between the posterior pulmonary valve cusps. There is good evidence, however, that the real line of fusion, while terminating in the same manner, otherwise lies further to the right, as has been illustrated previously[15, 17] and pointed out by Goor, Edwards, and Lillehei.[2] Occasionally, in anomalous hearts and even in normal specimens (Fig. 4), both an actually fibrous raphe and a groove are present, and the pattern of arterial blood supply of the crista supraventricularis is in agreement with this. The groove probably represents the furthest advance of the process of undermining of the conus septum which contributes, as will be shown later, to the formation of the tricuspid valve.

Saphir and Lev, in 1941,[12] employed the term "parietal and septal muscle bundles" for Keith's infundibular bands, again without mentioning the term crista supraventricularis. By 1953 Lev[8] had changed the names slightly to "parietal and septal bands" which together were then said to form the crista supraventricularis. Neither of the two bands is named in either the *B.N.A.* or *N.A.*

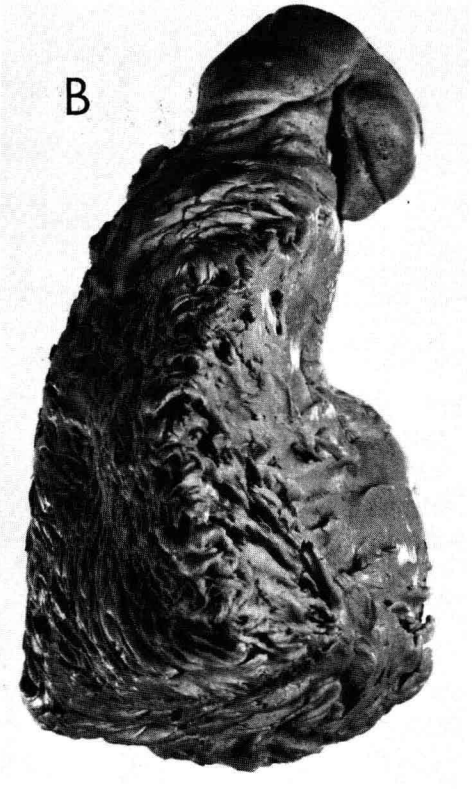

FIG. 1*B*

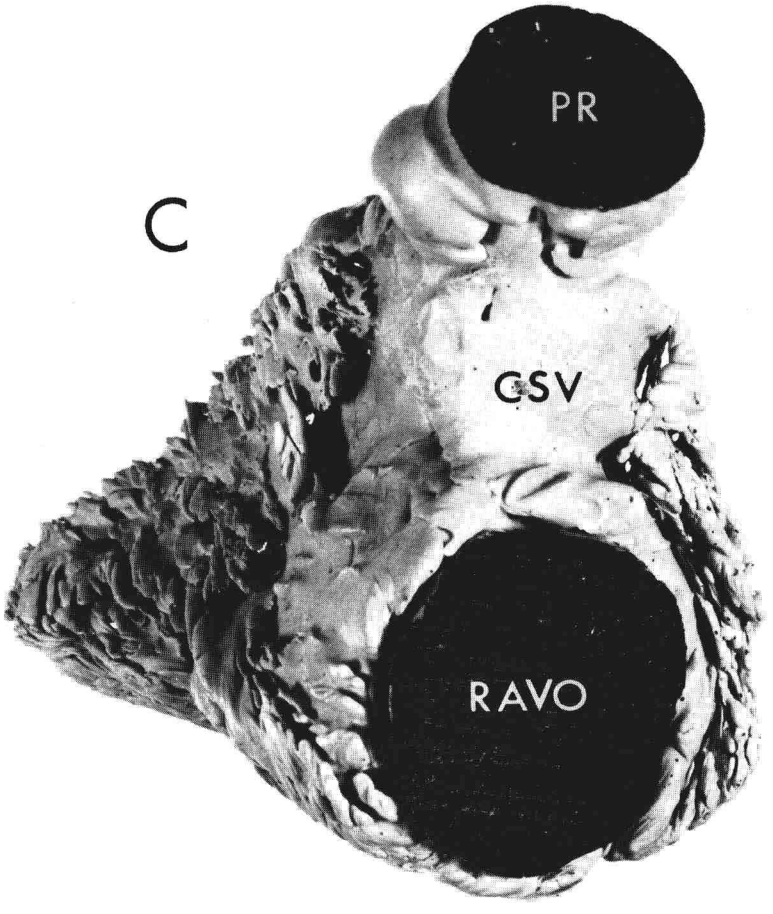

FIG. 1C

Kjellberg, Mannheimer, Rudhe, and Jonsson,[6] in their illustrations, used the term "crista supraventricularis" in more or less the original sense and applied the terms "parietal" and "septal" bands, respectively, to a lateral extension of the crista running along the roof of the right ventricle towards the acute margin of the heart and another extension which runs in an apical direction along the right side of the ventricular septum (Fig. 5). Lev and Eckner, in 1966,[9] equated the parietal band with the crista supraventricularis while the septal band apparently was not felt to be part of the crista but was now, together with the moderator band, considered to be identical with the trabecular septomarginalis, employing the latter term in a sense rather different from that used by Tandler,[14] the originator of the term. Lev and Eckner may have been influenced by the fact that in ungulates, *e.g.*, the pig, the septal band is separate from the septum, and together with the moderator band, forms a muscle bundle which originates high

on the septum and traverses the lumen of the right ventricle to terminate on the parietal right ventricular wall.

Van Praagh[18] not only equates the parietal band with the crista supraventricularis as did Lev and Eckner, but recently introduced a third term, "distal conus septum," for the same (adult) structure. Similarly, "proximal conus septum" is used synonymously with septal band. The use of the embryological term "conus septum" for an adult, *i.e.*, fully developed, anatomical structure cannot be recommended. Moreover, the use of the adjectives "proximal" and "distal" is rather puzzling since the terms proximal and distal conus septum are not known in cardiac embryology. Some authors speak of a conus septum and a truncus septum (or truncoconal septum if taken together),[1, 7, 11, 15, 17] while others, *e.g.*, Tandler,[14] used the terms proximal and distal bulbus septum for the corresponding structures in the embryo heart. Introduction of the term conus septum as an adult anatomical term would tend to confuse further an already confused state of affairs.

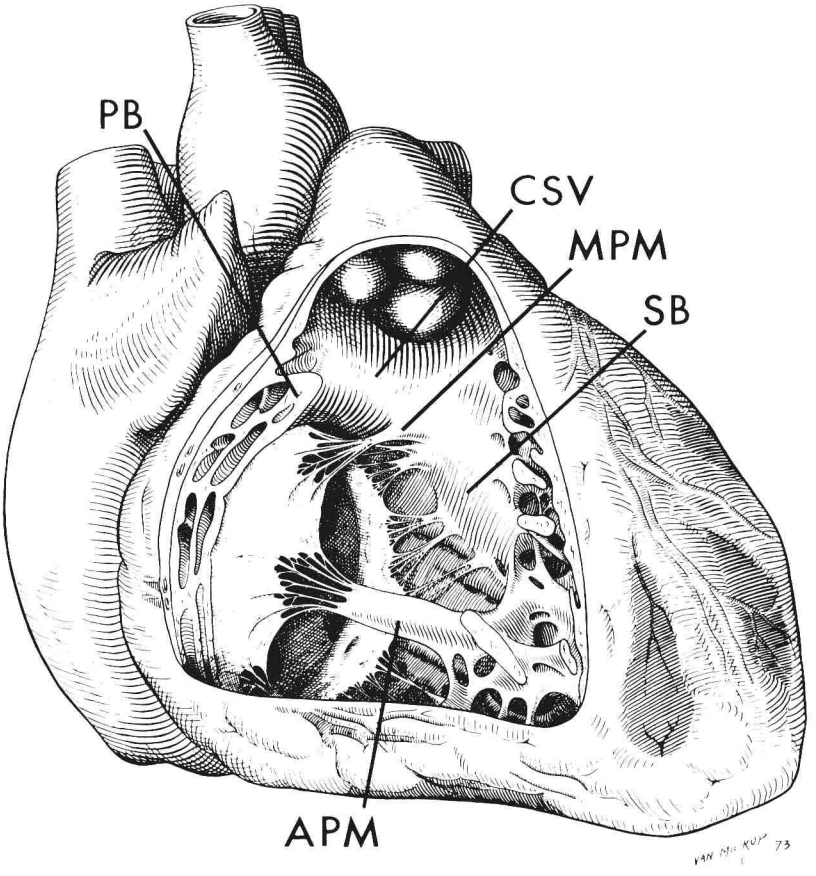

FIG. 2. Interior of right ventricle. *PB*, parietal band. *CSV*, crista supraventricularis. *MPM*, medial papillary muscle. *SB*, septal band. *APM*, anterior papillary muscle.

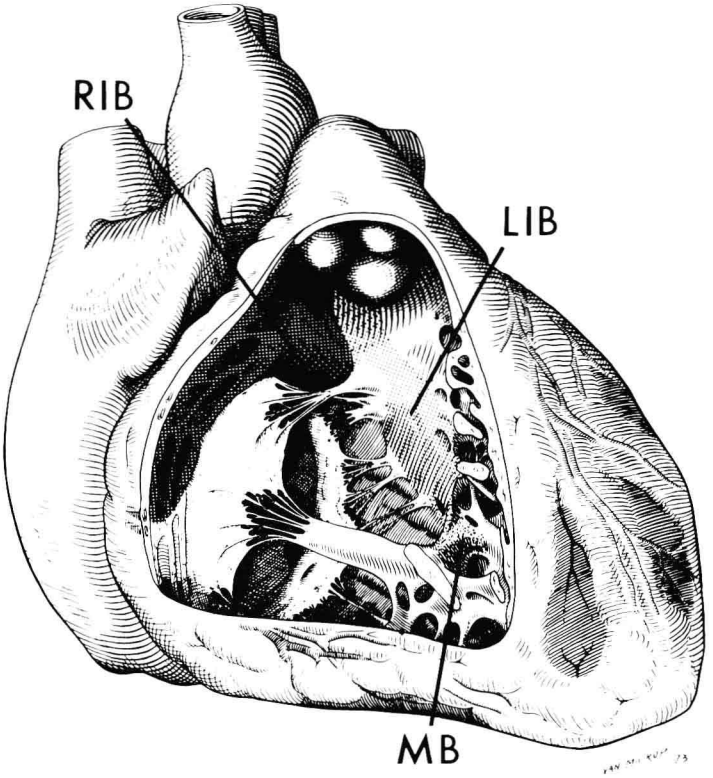

FIG. 3. *RIB*, right infundibular band. *LIB*, left infundibular band. *MB*, moderator band. (Nomenclature according to Keith.[4])

Towards the apical portion of the ventricle a variable, but usually well developed and quite stout, muscle band bridges the lumen of the right ventricle (Fig. 2). This large trabecula takes its origin from the ventricular septum at the inferior termination of the septal band (with which it is continuous) and loses itself among the trabeculae carneae of the acute margin of the right ventricle. Hence, the name trabecula septomarginalis introduced by Tandler,[14] who notes that both Leonardo da Vinci and Vesalius apparently had illustrated it. The *N.A.* of 1966 also lists the term "trabecula septomarginalis" but defines it as connecting the ventricular septum to the base of the anterior papillary muscle, which is a slightly different usage than that employed by Tandler, and further states that "it was formally known as the moderator band." The term "moderator band" originated with King,[5] who called it the "moderator band of distention." King believed that it prevented excessive dilation of the right ventricle during diastole which, if not checked, would displace the anterior papillary muscle of the tricuspid valve so far from the septum that function of the tricuspid valve would be interfered with, presumably causing incompetence. King's view must be incorrect since in many animals the moderator band consists only of fibers of the conduction system and would be much too feeble to exert any restraining action. Moreover, in some human hearts it is very thin or even absent

without resulting in any dysfunction of the tricuspid valve apparatus. Tandler[14] postulated that the trabecula septomarginalis phylogenetically is homologous to the right portion of the muscular ridge found in the reptilian heart, and believed it to be a rudiment thereof. Occasionally, particularly in anomalous human hearts, the septal band may be more or less completely separated off the ventricular septum and, with the trabecula septomarginalis, forms a large muscle bundle which crosses the ventricular lumen originating near the septal termination of the crista supraventricularis as is normally seen in the pig heart.

Because, as will become apparent later, the nomenclature as used by Kjellberg *et al.* makes much sense from a developmental point of view, I have adopted it.[15, 17] The central and most prominent part of the inverted U-shaped muscular band complex which separates the inflow and outflow portions of the right ventricle is therefore called the crista supraventricularis. "Parietal band" is used for the right limb of the U and "septal band" for the left limb. The three components have an entirely different embryological origin; they just happen to become lined up in normal development but do not necessarily do so in anomalous hearts.

A number of papillary muscles anchor the tricuspid valve cusps to the right

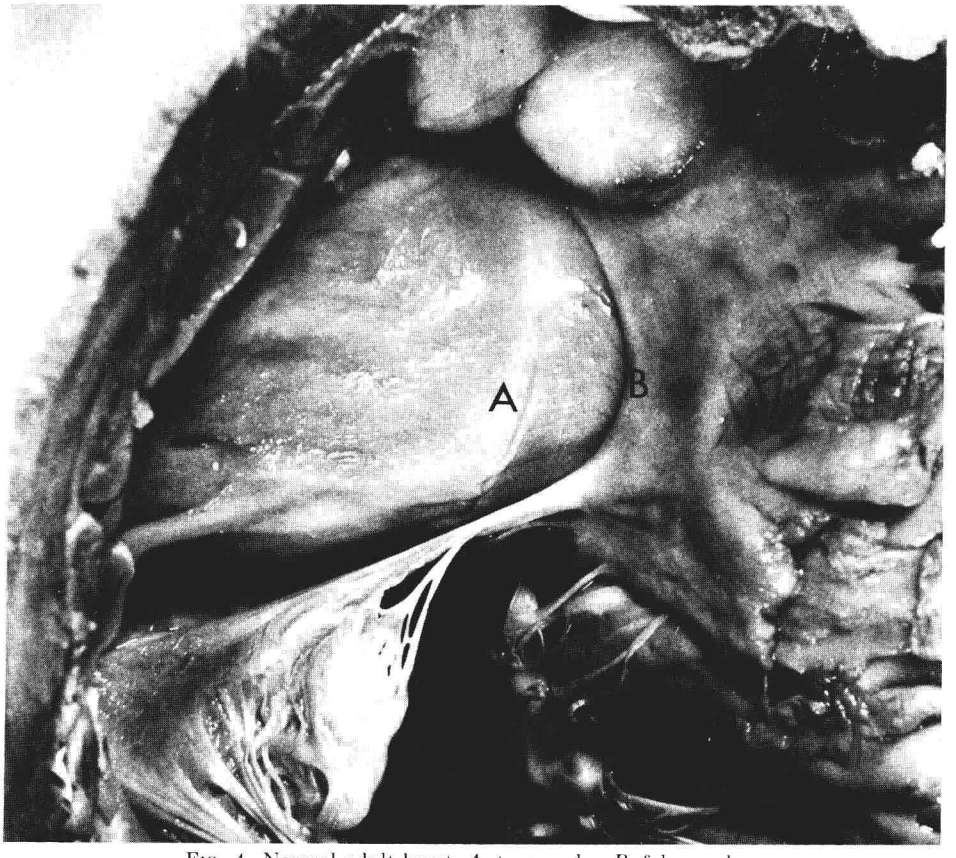

FIG. 4. Normal adult heart. *A*, true raphe. *B*, false raphe.

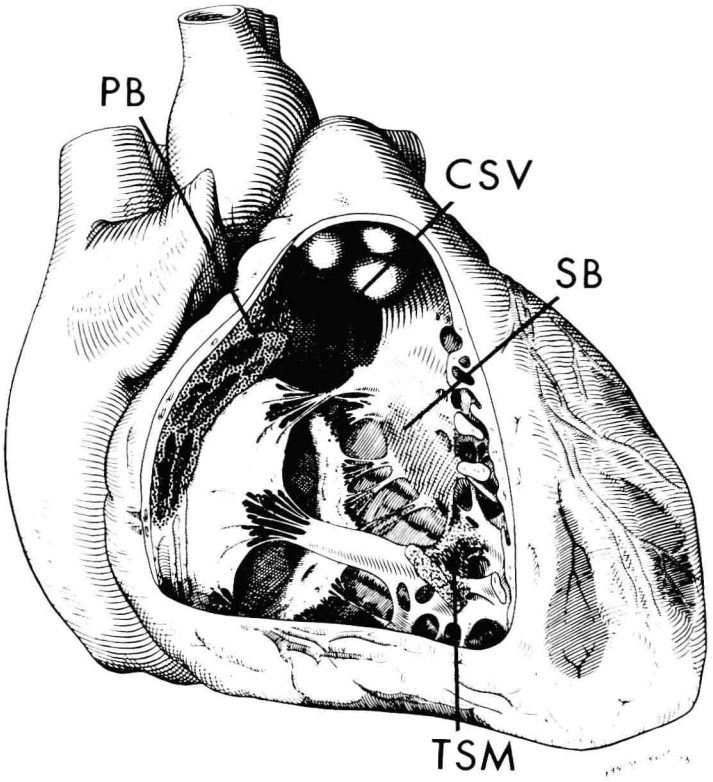

FIG. 5. *PB*, parietal band. *CSV*, crista supraventricularis. *SB*, septal band. *TSM*, trabecula septomarginalis (Tandler[14]). (Nomenclature according to Kjellberg *et al.*[6])

ventricular wall by means of a large number of slender fibrous strands called chordae tendineae. Two of these papillary muscles, the medial or conal papillary muscle, already referred to above, and the anterior papillary muscle, are reasonably constant in position though variable in size. The medial or conal papillary muscle (also referred to sometimes as the muscle of Lancisi) originates from the ventricular septum approximately where the crista supraventricularis joins the septal band. While always small, it is a distinct structure in the newborn heart but it tends to become less prominent with age and in some adult specimens may be reduced to a small tendinous patch. It receives chordae tendineae from the medial portion of the anterior cusp of the tricuspid valve and also often a few from the most anterior portion of the septal cusp. A variable number, usually two or three, very small papillary muscles originate from the posterior border of the septal band and receive chordae tendineae from the anterior half of the septal cusp. The uppermost of these small papillary muscles is located closely behind the medial papillary muscle. In congenital cardiac anomalies in which the crista supraventricularis is grossly abnormal, *e.g.*, tetralogy of Fallot, the medial papillary muscle is absent and its function is taken over by this uppermost septal papillary muscle which is then often mistakenly interpreted to be the medial papillary muscle.

The usually well developed and quite large anterior papillary muscle of the right ventricle in man originates from the outer, lateral one-third of the trabecula marginalis. In the intact heart, this point of origin may be projected on the anterior wall of the right ventricle in the manner illustrated in Figure 6. Thus, damage to the anterior papillary muscle may be avoided in carrying out a right ventriculotomy at the time of surgery. The anterior papillary muscle usually receives chordae tendineae from both the lateral portion of the anterior tricuspid valve cusp and the posterolateral cusp. The remainder of the chordae tendineae of the posterolateral and septal cusps insert onto a variable number of small papillary muscles originating from the posterior part of the ventricular septum and inferior or diaphragmatic wall of the right ventricle.

It is not uncommon for the posterolateral cusp to be subdivided into two smaller cusps, thus creating a quadricuspid valve. Occasionally, the small portion of the septal cusp normally overlying the membranous part of the interventricular septum is adherent to it, thus creating a small gap in the septal cusp. This is of no functional significance and ordinarily does not cause incompetence of the valve.

EMBRYOLOGY

In an embryo of about 6 mm crown-rump (C-R) length, the heart on external

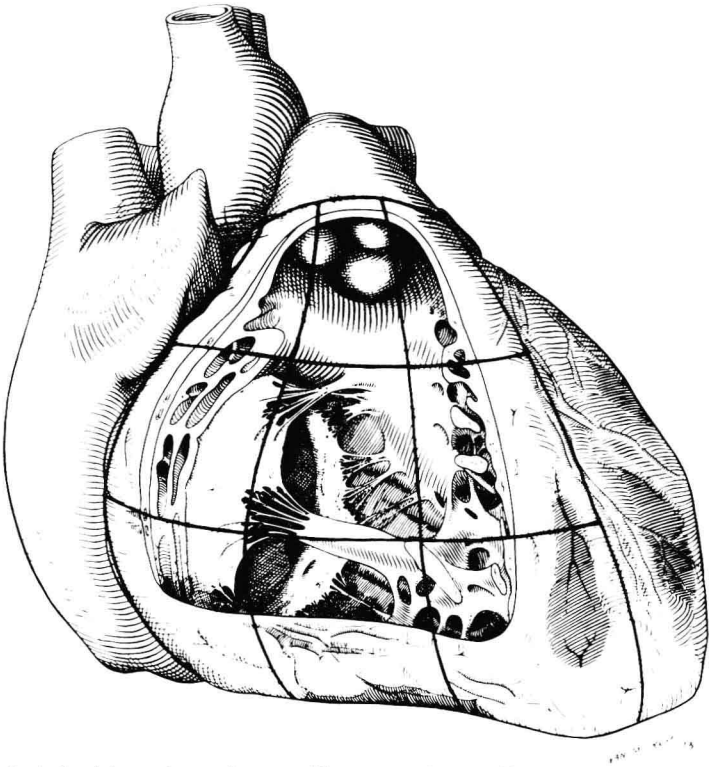

FIG. 6. Projected origin and attachment of large anterior papillary muscle of right ventricle. (See explanation in text.)