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The Univentricular Atrioventricular Connections

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= Abstract = The authors examined nine autopsied hearts with univentricular atrioventricular connection (UAVC), and described the important pathologic findings of each cardiac segment. The classification of our cases was based on the morphology of main chamber and the status of atrioventricular connection. Left ventricular type was seen in five cases; one case of double inlet, three cases of absent right atrioventricular connection and one absent left atrioventricular connection. Of four cases of right ventricular type, two were double inlet, one absent right atrioventricular connection and one absent left connection. The pattern of the apical trabeculation of main chamber, the relationship between the septum and the crux cordis, and the presence and position of rudimentary chamber were the essence in determining the type of UAVC. The distribution pattern of delimiting arteries may be used as a guide to determine the ventricular loop when the anatomical position of the rudimentary chamber is equivocal. In the cases of absent atrioventricular connection the ventricular loop can be determined by the ventricular type of the main chamber and the site of absent connection.

Key Words: ***Univentricular atrioventricular connection (UAVC), Univentricular heart, Single ventricle, Ventricular loop, Delimiting artery***

INTRODUCTION

The heart with one main chamber is an uncommon anomaly, having an incidence of 0.7 to 1.0 percent of all congenital heart diseases (Rowe *et al.* 1981; Hong *et al.* 1983). This anomaly has been the source of increased interest and controversy in the embryological, pathological and surgical aspects. Various terms of 'single or common ventricle' (Van Praagh *et al.* 1964), 'primitive ventricle' (Lev *et al.* 1969), 'double inlet ventricle' (de la Cruz and Miller 1968; Munoz-Castellanos *et al.* 1972), 'univentricular heart' (Anderson *et al.* 1976) and 'univentricular atrioventricular connection (UAVC)' (Anderson *et al.* 1983; Anderson *et al.* 1984) have been applied and the embryologic explanation and diagnostic criteria are different by the nomenclatures. The classification of this anomaly is based on the morphologic type of the main chamber and the status of atrioventicular connection and it is important in the surgical standpoint, because the cardiac conduction system and the distribution of coronary arteries are closely related to the morphologic class (Ahnn *et al.* 1983; Park *et al.* 1984; Stefanelli *et al.* 1984). Step-by-step analysis of each cardiac segment is helpful in the study of complex cardiac anomalies including UAVC. But clinical analysis of the cardiac segments is often insufficient or equivocal, and the knowledge on the degree of importance and the interrelation of the findings based on autopsy specimen examination is valuable in those cases.

The authors examined nine autopsied hearts with UAVC and described the important pathologic findings of each cardiac segments in the morphologic classes and their implications for clinical diagnosis.

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Table 1. Identification of the cases

Case	Autopsy No.	Diagnosis	Age/Sex	Segmental Dx.	Remark
1	HYUH A85-7	UVH of LV type double inlet	7m/F	TGA (S,D,S)	
2	CMC A79-42	UVH of LV type absent Rt AVC	4m/F	NRGA (S,D,S)	
3	SNUH A82-86	UVH of LV type absent Rt AVC	3d/M	TGA (S,D,D)	subarachnoid hemorrhage
4	SNUH A83-119	UVH of LV type absent Rt AVC	1d/F	NRGA (S,D,S)	lung hemorrhage
5	SNUH A80-33	UVH of LV type absent Lt AVC	3m/M	ACM (S,L,D)	Rt juxtaposition of atrial appendages
6	SNUH A78-21	UVH of RV type double inlet	16y/M	DORV (A,D,D)	situs inversus levocardia
7	SNUH A82-26	UVH of RV type double inlet	1d/M	TA (S,D,X)	cong. lymphangiectasia ear malformation
8	SNUH A81-62	UVH of RV type absent Lt AVC	15d/M	DORV (S,D,S)	imperforate anus
9	SNUH A85-55	UVH of RV type absent Rt AVC	1m/M	TGA (A,L,X)	asplenia

HYUH: Hanyang University Hospital
SNUH: Seoul National University Hospital
UVH: univentricular heart,
TGA: transposition of great arteries,
ACM: anatomically corrected malposition,
TA: truncus arteriosus
CMC: Catholic Medical College
AVC: atrioventricular connection,
NRGA: normally related great arteries,
DORV: double outlet right ventricle,

MATERIALS AND METHODS

Nine heart specimens in which there were single functioning ventricular chamber with double or single inlet were examined. Seven cases were from the Heart Collection of Department of Pathology, College of Medicine, Seoul National University. Others were from Catholic Medical College and Hanyang University Hospital (Table 1).

Segmental analysis was applied in examining the atrial morphology, atrioventricular connection, atrioventricular valves, ventricular septum, main and rudimentary chamber, coronary arteries and ventriculoarterial relation.

Definitions and Diagnostic Criteria: The univentricular atrioventricular connection (UAVC) is defined as a collective term for those hearts in which the atria connect to only one ventricle (Anderson *et al.* 1984). The main chamber may have the morphologic features of right, left or indeterminate ventricle. The atrioventricular connection may be double inlet (two separate valves or common valve), absent right atrioventricular connection or absent left atrioventricular connection. The atrioventricular valves are termed as either right or left atrioventricular valve rather than tricuspid or mitral valves, because they frequently have unusual features and can not be designated as normal mitral or tricuspid valves (Tynan *et al.* 1979). Three types of main ventricular chamber and three types of atrioventricular connection were the main defining features of nine morphologic classes of UAVC. The rudimentary chamber may have its arterial outflow (outlet chamber) or not (trabecular pouch). The crux cordis is the posterior crossing point of the transverse atrioventricular groove and the vertical septation of the atria. The descending coronary arterial branches at the external junction between the main and rudimentary chambers are called left and right delimiting arteries (Lev *et al.* 1969).

RESULTS

The summary of the segmental observation of the cases and the associated anomalies are listed in Table 2.

A. Univentricular Heart of Left Ventricular Type

1) Double Inlet (Case 1)

The atria were connected to the main chamber of left ventricular type through two separate valves. The right atrioventricular valve had three cusps, and two distinct and one small papillary muscles were seen. The left atrioventricular valve had two cusps and more than three small papillary muscles were attached to the valve. There was a posteromedian muscle ridge (PMMR) between the posterior attachments of two atrioventricular valves. The posterior papillary muscle of right atrioventricular valve was connected to the PMMR at its apical end. The head of the PMMR was connected to the crux cordis (Fig. 1). The septum between the main and rudimentary chambers was not connected to the crux and a restrictive outlet foramen was seen at the upper portion of the septum. The trabeculation of the main chamber was left ventricular type and the left atrioventricular valve and the pulmonic valve were in fibrous continuity. The rudimentary chamber was an anterior left outlet chamber ascending to aorta in right upper portion. The aorta was in the left anterior of pulmonic valve. Single coronary artery arising from right aortic sinus gave rise to left and right circumflex arteries and a descending branch, the right delimiting artery. The left delimiting artery was not conspicuous.

2) Absent Right Atrioventricular Connection (Cases 2,3 and 4)

The right atrioventricular connection was absent and the atrial outflow was connected to the main chamber of left ventricular type through the left atrioventricular valve. A small dimple noticed in the muscular floor of the right atrium was connected to the main chamber (Fig. 2). The lateral junction between the right atrium and the ventricular mass was presented as a deeply excavated atrioventricular groove. The right posterior shoulder of the ventricle was also depressed (Fig. 3A). The left atrioventricular valve had two cusps with papillary muscle attachments. The main chamber of left ventricular type had fine trabeculae and the rudimentary chamber in the right anterior was an outlet chamber ascending left superiorly. Small portion of the outlet chamber had definite trabeculae of the right ventricular type (Fig. 3B). The septum between the main and rudimentary chambers was not connected to the crux. There was an outlet foramen in cases 2 and 3, but none found in case 4. The aorta was situated right and posterior and arose from the main chamber in cases 2 and 4, and the pulmonary artery from the rudimentary chamber. The ventriculoarterial connection was discordant and aorta was right anterior in case 3. Two aberrant coronary arteries were seen in front of the infundibulum and pulmonary artery in case 4. The anterior descending coronary artery arising from left coronary artery was identified as the left delimiting artery. The right delimiting artery was inconspicuous.

3) Absent Left Atrioventricular Connection (Case 5)

The left atrioventricular connection was absent and the atrial outflow was connected to the main chamber of left ventricular type through the right atrioventricular valve. The left atrial auricle was juxtaposed between the right atrial auricle and the great vessels (Fig. 4). The right atrioventricular valve had two cusps with several papillary muscle attachments. The main chamber of left ventricular type had fine trabeculae and the rudimentary chamber was an outlet chamber in the left anterior ascending posterosuperiorly. The trabecular septum did not extend to the crux and a large outlet foramen was seen. The main chamber was connected to the aorta in the right anterior and the rudimentary chamber to the pulmonary artery. The right delimiting artery was a descending branch of the right coronary artery. The left delimiting artery was a branch of the left circumflex artery similar to the marginal artery.

B. Univentricular Heart of Right Ventricular Type

1) Double Inlet (Cases 6 and 7)

The atria were connected to the main chamber of right ventricular type through a common valve. The main chamber had coarse trabeculae and branching muscular trunk similar to the trabecula septomarginalis of the right ventricle (Fig. 5). The rudimentary chamber was a slit-like trabecular pouch in the left posterior wall. The septum forming the posterior wall of the main chamber was connected to the crux. The connection between the two chambers was made through a ventricular septal defect of atrioventricular septal defect type presenting as the only opening of the rudimentary chamber. The ventriculoarterial connection was a

Table 2. Summary of the segmental observation and the associated anomalies of the cases

Case	No. Diagnosis	Atrium	A-V connection	Main ch.	Rudim. ch.	V-A connection	Associated anomalies
1	UVH of LV type double inlet	S. solitus ASD II	R: 3 cusps L: 2 cusps	LV type PMMR	Lt-ant (l-loop) Outlet chamber	Discordant L-TGA	single coronary artery
2	UVH of LV type absent Rt AVC	S. solitus PFO	R: absent L: 2 cusps	LV type	Rt-ant (d-loop) Outlet chamber	Concordant NRGA	
3	UVH of LV type absent Rt AVC	S. solitus ASD II	R: absent L: 2 cusps	LV type	Rt-ant (d-loop) Outlet chamber	Discordant D-TGA	preductal coarctation PDA
4	UVH of LV type absent Rt AVC	S. solitus ASD II	R: absent L: 2 cusps	LV type	Rt-ant (d-loop) Outlet chamber	Concordant NRGA	aberrant coronary artery closed outlet foramen
5	UVH of LV type absent Lt AVC	S. solitus ASD II Juxtaposition	R: 2 cusps L: absent	LV type	Lt-ant (l-loop) Outlet chamber	Concordant D-TGA	
6	UVH of RV type double inlet	RA isomerism Common atrium	Common valve	RV type	Lt-post (d-loop) Trabecular pouch	DORV D-TGA	isolated levocardia
7	UVH of RV type double inlet	S. solitus Common atrium	Common valve	RV type	Lt-post (d-loop) Trabecular pouch	Truncus arteriosus	
8	UVH of RV type absent Lt AVC	S. solitus Single RA auricle	R: 3 cusps L: absent	RV type	Lt-post (d-loop) Trabecular pouch	DORV NRGA	aortic arch interruption
9	UVH of RV type absent Rt AVC	RA isomerism ASD I+SV	R: absent L: 3 cusps	RV type	Rt-post (l-loop) Trabecular pouch	Pulm. atresia	asplenia TAPVR-supracardiac

ASD II: atrial septal defect secundum type
ASD I: atrial septal defect primum type
ASD SV: atrial septal defect sinus venosus type
PFO: patent foramen ovale
PDA: patent ductus arteriosus
TAPVR: total anomalous pulmonary venous return
PMMR: posteromedian muscle ridge

double outlet from the right ventricular main chamber and the aorta was right anterior in case 5 and truncus arteriosus in case 6. The left delimiting artery descended from the left coronary artery and the right delimiting artery was the posterior descending artery arising from the crux.

2) Absent Left Atrioventricular Connection (Case 8)

The left atrioventricular connection was absent and the atrial outflow was connected to the main chamber of right ventricular type through the right atrioventricular valve. The right atrioventricular valve had three cusps and three papillary muscle attachments. The main chamber had coarse trabeculae and the rudimentary chamber was represented as a slit-like trabecular pouch with no opening. The septum between the two chambers was connected to the crux. The two great arteries arose from the main chamber; the aorta was in the right posterior of the pulmonary artery. The descending branch of the left coronary artery formed the left delimiting artery and the posterior descending artery at the crux, the right delimitng artery (Fig. 6).

3) Absent Right Atrioventricular Connection (Case 9)

The right atrioventricular connection was absent and the atrial outflow was connected to the main chamber of right ventricular type through left atrioventricular valve. The lateral border of right atrioventricular groove was deeply excavated and the right posterior shoulder was depressed. The left atrioventricular valve had three cusps and three papillary muscle attachments. The main chamber had coarse trabeculae and a right posterior trabecular pouch was recognized as the rudimentary chamber which had considerable space and fine trabeculae (Fig. 7). The septum between the two chambers was connected to the crux. The only communication noticeable between the two chambers was a muscular ventricular septal defect at its right anterior margin. The main chamber had single aortic outlet. The pulmonary artery was atretic and the pulmonary flow was through the ductus arteriosus. The descending branch of the right coronary artery was the right delimiting artery and the posterior descending artery, the left delimiting artery.

DISCUSSION

The heart with one effective pumping chamber has been well defined pathologically for many years, but recently its definition, nomenclature, and classification have generated many controversies and polemics. Van Praagh *et al.* (1964) had defined a single or common ventricle as one ventricular chamber which receives both the tricuspid and mitral valves, or a common atrioventricular valve, and classified such ventricle into four types. This definition excluded tricuspid and mitral atresia. Lev *et al.* (1969) used te term single primitive ventricle. De la Cruz and Miller (1968) and Munoz-Castellanos *et al.* (1973) used the term double inlet ventricle and reported left ventricular type and right ventricular type with comments on embryology. Anderson *et al.* (1976) used the term univentricular heart and newly redesigned a generic term 'univentricular atrioventricular connection (UAVC)' (Anderson *et al.* 1983). Under this name they included hearts with double-inlet, absent right, or absent left atrioventricular connection, that share a common finding of only one of their ventricular chambers connected to the atria. Also, since each type of the univentricular atrioventricular connection can coexist with any of three ventricular morphologies, left ventricular type, right ventricular type, and indeterminate type, a total of nine types are classified.

In segmental analysis of the hearts with univentricular atrioventricular connection the atria can be arranged in situs solitus, inversus, or ambiguus (Becker & Anderson 1981a). Mostly, the atrial situs is solitus, but isomerism is frequently found in right ventricular type and indeterminate type. In our series solitus was in seven cases, right atrial isomerism in two. Both cases of isomerism were of the right ventricular type. There are always various types of atrial septal defects and frequent associations with other anomalies. In our series all specimens had interatrial communications and one (case 5) had an uncommon anomaly, i.e. the right juxtaposition of atrial appendages (Melhuish & Van Praagh 1968).

In double inlet atrioventricular connection with separate atrioventricular valves, the atrioventricular valves are both usually similar in pattern to mitral valve, and particularly prone to malformations themselves (Quero-Jimenez *et al.* 1979). In our one specimen with double inlet atrioventricular connection (case 1) right atrioventricular valve had three leaflets, but one of them was very hypoplastic, and the arrangement of papillary muscle attachments were abnormal. In double inlet atrioventricular connection with a common valve

pattern, all valves had the leaflet pattern of the common valve, although usually four leaflets were recognizable. The attachment pattern of the papillary muscles were similar to that of the common valve (Keeton *et al.* 1979b). The common valve was found more frequently in right ventricular type than in left ventricular type. In our series the common valve was seen in two cases, both of which were of right ventricular type.

When one atrioventricular connection was absent, the floor of the blind-ending atrium was muscular and since it was separated from the underlying ventricular mass by the atrioventricular sulcus, the atrium could be lifted easily from the underlying ventricular mass (Becker & Anderson 1981b). We also observed characteristic features of a deeply excavated atrioventricular groove of absent connection and a depressed ventricular shoulder on the same side (Fig. 3A). Some argued that tricuspid atresia is not a univentricular heart (Bharati & Lev 1979), but when analyzed in terms of ventricular morphology it is inescapable that the hearts with double inlet to a left ventricular chamber or with absence of right atrioventricular connection in the setting of a main left ventricular chamber are directly comparable (Anderson *et al.* 1979). When the right connection is absent, in the floor of the right atrium the dimple frequently seen overlies the atrioventricular membranous septal component of the central fibrous body (Anderson *et al.* 1977), and which considered by some to represent the site of the tricuspid valve at one time. Either when probed with a pin or transilluminated it can be shown to connect with the left ventricular chamber. We also confirmed this (Fig. 2). The situs of the atrioventricular valves corresponded to that of the ventricles of entry, not to that of the atria of exit. When one atrioventricular connection is absent, the number and morphology of the leaflet of the another connecting atrioventricular valve corresponded with those of the ventricular morphology of the main chamber.

The pattern of the apical trabecular component of the main chamber and the relationship between the septum and the crux cordis are the essence in the determination of type of the hearts with univentricular atrioventricular connection (Anderson *et al.* 1983). Whether there is double inlet, absent left, or right atrioventricular connection, the basic ventricular morphology and the relationship between the septum and the crux are the same. In left ventricular type the atria connect to the left ventricular main chamber posteriorly to an interventricular septum which does not extend to the crux. In right ventricular type the atria connect to the right ventricular main chamber anteriorly to an interventricular septum which almost always extends to the crux. In indeterminate type, which we have not experienced in this study, the atria connect to a solitary ventricle that is present with neither right nor left trabecular patterns. The univentricular heart of the indeterminate type is quite distinct from a huge ventricular septal defect, the so-called common ventricle or single ventricle of type C of Van Praagh (1964). In the latter anomaly the trabecular septum, although hypoplastic, definitely separates the right and left trabecular zones and there is usually a hyoplastic inlet septum.

In double inlet univentricular heart with separate atrioventricular valves they are usually separated the inlet part of the main chamber by a prominent posterior ridge, termed posteromedian muscle ridge (Devloo-Blancquaert *et al.* 1978). It can be found frequently, as in our case 1, where malalignment exists between the ventricular septum and the junction of both atrioventricular valve rings. It has been considered embryologically to be created by the two inflow tracts, entering the primitive ventricle. The cranial insertion of the posteromedian muscle ridge is always situated in the crux cordis.

Since it has been shown that this ridge does not carry any conduction tissue in double inlet univentricular heart of left ventricular type, the ridge is a safe ideal site for the septation procedure (Anderson *et al.* 1974).

Chambers which do not possess any inlet portions are not ventricles and are described as rudimentary chambers (Tynan *et al.* 1979). They may be of two varieties; outlet chambers which possess both trabecular and outlet portions or trabecular pouches having only a trabecular part. They may be of right or left ventricular type depending upon the nature of their trabecular portion and are the converse type of the main chamber. Univentricular hearts of left ventricular type usually have an outlet chamber, whereas those of right ventricular type usually have a trabecular pouch. In our series all cases of left ventricular type (cases 1-5) had outlet chambers and those of right ventricular type (cases 6-9) had trabecular pouches.

The size of rudimentary chamber is variable. In left ventricular type the rudimentary chamber always possesses not only an infundibular portion (Van Praagh *et al.* 1983) but also a trabecular

Table 3. Morphologic differences between the left and right ventricular types in univentricular atrioventricular connections

Segmental description	LV type	RV type
Atrial situs	Solitus	Isomerism frequent
Atrioventricular value		Common valve frequent
Trabeculae of main chamber*	Fine	Coarse
Position of rudimentary chamber*	Anterior to septum	Posterior to septum
Character of rudimentary chamber	Outlet chamber	Trabecular pouch
Septum-crux connection*	Present	Absent
Ventriculoarterial connection	Discordant or concordant	Double or single outlet
Prominent coronary artery	Anterior descending coronary artery	Posterior descending coronary artery

*: Pathognomonic features

componet of the right ventricular type which is usually hypoplastic. In right ventricular type the rudimentary chamber also shows variable size from a well-defined hypoplastic chamber to a tiny slit-like structure that may resemble a left ventricular trabeculation (Shinebourne *et al.* 1980). In our series the rudimentary chambers were also of varying size. The presence and position of the rudimentary chamber were important in determining ventricular type and ventricular loop and were also related to the course of the non-branching bundle, especially in double inlet left ventricular type (Wenink 1978). In left ventricular type the two major positions of the rudimentary right ventricular chamber were anterosuperior on the right shoulder of the main chamber (normally related; d-loop) or on the left shoulder (inverted; l-loop), and they were found in approximately equal frequencies. In right ventricular type the rudimentary left ventricular chamber was always positioned posteroinferiorly within the ventricular mass. Soto *et al.* (1982) also documented these findings angiographically.

We considered that the ventricular loop is easily determined by the ventricular morphology of the main chamber and the site of absent connection when one atrioventricular connection is absent. In cases of absent right atrioventricular connection of left ventricular type (cases 2,3 and 4) the rudimentary chamber was normally related (d-loop), and in absent left connection (case 5), inverted (l-loop). When the main chamber was right ventricular type, it was normally related (d-loop) in absent left atrioventricular connection (case 8), and inverted (l-loop) in absent right connection (case 9).

The terms right and left delimiting arteries are usually applied to the epicardial coronary vessels in UAVC (Lev *et al.* 1969), but the term anterior descending coronary artery can also be applied to one of the vessels with the following criteria: a more prominent delimiting artery arising early at the coronary arterial stump and arising in an obtuse angle. The identification of the distribution of the delimiting arteries may be used as a guide to determine the ventricular loop (Van Praagh 1972) especially when the anatomical position of the rudimentary chamber is equivocal (Keeton *et al.* 1979a). The schematic representation of the disposition of the delimiting arteries in each type of univentricular heart is in Fig. 8.

Most of the ventriculoarterial connections of the double inlet univentricular heart of left ventricular type are usually discordant and obstruction of one or the other great artery is frequent (Becker & Anderson 1981a & b). When the ventriculoarterial connection is concordant in this type of heart, however, it is frequently called the 'Holmes heart', commonly characterized by an elongated infundibulum and pulmonary stenosis. When the right atrioventricular connection is absent, there is most often a ventriculoarterial concordance with normal arterial relationship as compared with the double inlet. When the left connection is absent, there is usually a discordance with the rudimentary chamber left-sided (inverted; l-loop). In univentricular heart of right ventricular type regardless of the type of atrioventricular connection, the ventriculoarterial connection most frequently encountered is double outlet from the main right ventricular chamber, usually with bilateral infundibulae, and next is sing-

le outlet. In our four cases of right ventricular type, two cases had double outlet, one truncus arteriosus, and one pulmonary atresia.

By this study we suggest the cardiac segments of special importance in UAVC to be the atrial situs, the atrioventricular connection, character of the main and rudimentary chambers, septum-crux relation, the ventriculoarterial connection and the coronary arteries, because they have significant morphologic differences between the left and right ventricular type (Table 3).

We endorse the opinion that all hearts can be categorized into one of two groups according to their atrioventricular connections (Anderson *et al.* 1984) In the first group, each atrial chamber is connected to its own ventricular chamber, *i.e.*, biventricular atrioventricular connection. As is well-known, normal hearts and most cases of congenital cardiac anomalies fall into this group. In the second group, the atria connect to only one ventricular chamber, *i.e.*, univentricular atrioventricular connection which has been the subject of our study.

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= 국문초록 =

단심실성 방실연결

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강순웅 · 서정욱* · 유시준** · 지제근* · 윤용수 · 홍창의 · 이정용*** · 박찬금**** · 이중달****

단심실성 방실연결은 단심실(single ventricle), 이중유입로 심실(double inlet ventricle) 등으로 불리우는 기형으로 정의 · 명칭 · 분류에 있어 많은 이견을 보이고 있다. 주심실의 형태와 방실연결상태에 따라 심장전도계, 관상동맥계등의 형태가 다르므로 분절식 분석(segmental analysis)에 의한 관찰이 특히 필요한 심기형이다.

저자들은 9예의 부검심장에서 단심실성 방실연결의 각 형별에 따른 형태학적 특징을 기술하고 검토하였다. 주심실은 좌심실형태가 5예, 우심실형태가 4예였으며, 이들의 방실연결 상태는 이중유입로인 경우가 각각 1예, 2예, 우측 방실연결이 없는 경우가 3예, 1예, 좌측 방실연결이 없는 경우가 각각 1예였다.

심방은 전예에서 중격결손을 동반하였고 2예는 대칭형 이었으며 이들을 모두 우심실형태의 주심실을 갖고 있었다. 이중유입로의 방실연결을 갖는 심장중 우심실형태는 모두 공통방실판막을 갖고 있었다. 주심실의 형태를 결정하는데는 심첨부 유주의 형태, 부심실의 존재여부 및 위치, 그리고 심실중격과 후측 교차점과의 관계가 중요한 지표가 되었다. 좌심실형태에서 부심실은 outlet chamber 형태였고, 우심실형태에서는 모두가 trabecular pouch형태였다. 심실환(ventricular loop)의 판단은 한쪽 방실연결이 없을때 주심실의 형태와 방실연결이 없는 쪽의 위치에 따라 결정되었으며, 특히 부심실의 해부학적 위치가 모호한 경우 경계동맥(delimiting artery)의 분포상태를 파악하는 것이 도움이 되었다. 좌심실형태에서는 전방 하행 관상동맥이 확실한 반면에 우심실형태에서는 후방 하행 관상동맥이 잘 발달되어 있는 것도 특징적으로 관찰되었다. 심실동맥연결은 좌심실형태에서는 정상(3예) 또는 전위(2예)였고, 우심실형태에서는 이중유출로(2예) 또는 단일유출로(2예)의 상태였다.

LEGENDS FOR FIGURES

Fig. 1. (case-1) Double inlet univentricular heart of left ventricular type. The trabeculae of main chamber (MC) is fine as in left ventricular type. The posteromedian muscle ridge (arrows) is seen between right atrioventricular valve (RAVV) and left atrioventricular valve (LAVV). Its cranial insertion is situated on the crux cordis(*), and its base is joined with posterior papillary muscle of RAVV. The trabecular septum does not extend to the crux cordis. AO =aorta, PA=pulmonary artery, OC=outlet chamber, OF=outlet foramen

Fig. 2. (case-2) Univentricular heart of left ventricular type, absent right atrioventricular connection. The floor of blind-ending right atrium is muscular and has a dimple (arrow). A pin (P) placed through the dimple passed into the left ventricular main chamber (MC). RA=right atrium, FO=fossa ovalis, OC=outlet chamber, LAVV= left atrioventricular valve

Fig. 3. (case-3) Univentricular heart of left ventricular type, absent right atrioventricular connection.
A: The right atrioventricular sulcus (AVS) is deeply excavated and the ventricular shoulder of the same side depressed (arrow), enabling the right atrium (RA) to be lifted easily from the underlying ventricular mass.
B: The internal view of this heart shows the trabecular pattern of main chamber (MC) and rudimentary chamber (RC). The rudimentary chamber is located anterior to the septum(*). The left atrioventricular valve (LAVV) has two leaflets and two papillary muscle attachments.

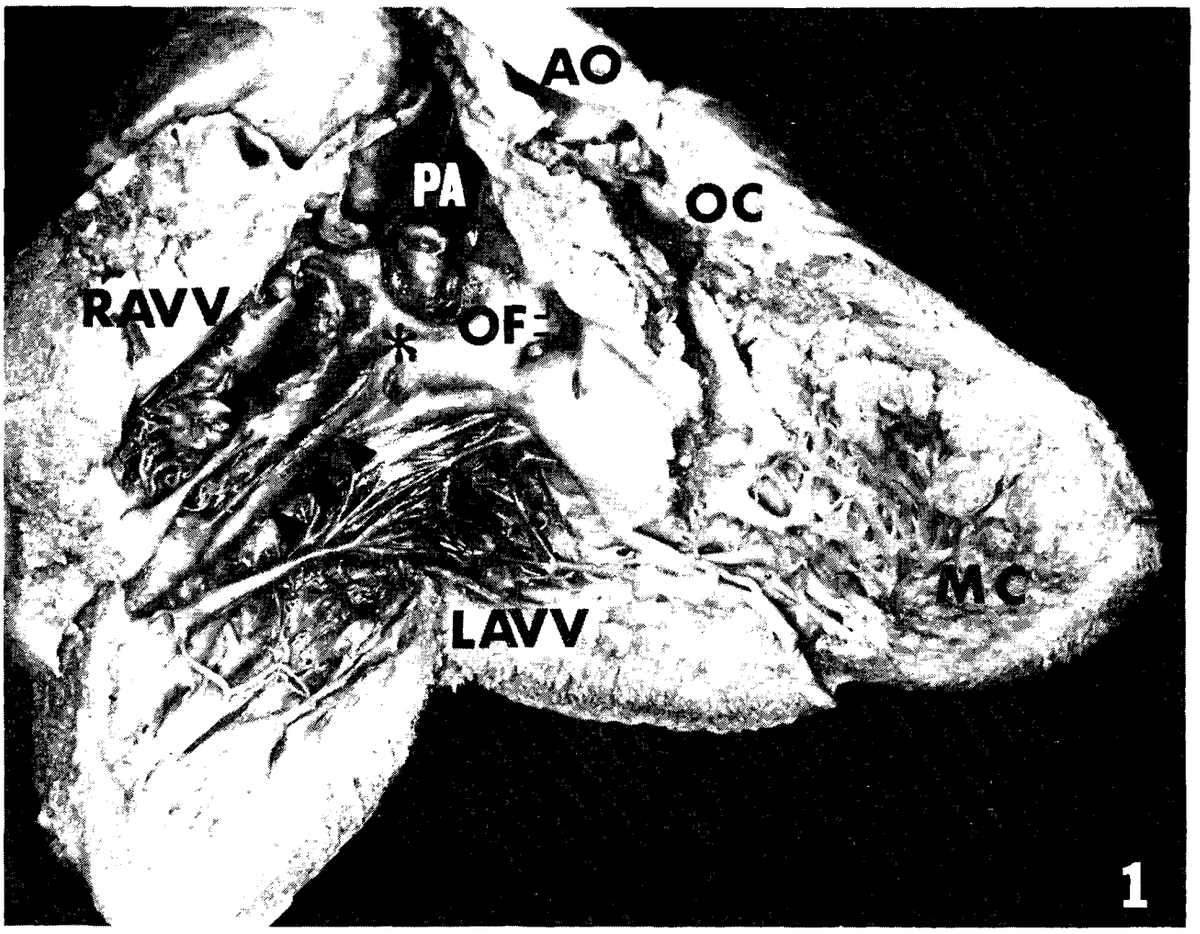
Fig. 4. (case-5) Univentricular heart of left ventricular type, absent left atrioventricular connection. This heart has a rare anomaly that the left atrial auricle (LAA) is juxtaposed between the right atrial auricle (RAA) and the aorta (AO); right juxtaposition of atrial appendages. The rudimentary chamber (RC) is located in the left anterior and its right delimiting artery is the anterior descending branch of the right coronary artery (arrows). The ventricular loop is inverted (I×loop). MC=main chamber, T=trachea

Fig. 5. (case-6) Double inlet univentricular heart of right ventricular type. The atria connect to the right ventricular main chamber (MC) through the common atrioventricular valve (CAVV). The main chamber has coarse trabeculae and branching muscle bundles similar to trabecula septomarginalis (TSM) of right ventricle. The common atrioventricular valve has a severe degree of straddling and partially connects to the rudimentary chamber through the atrioventricular septal defect (arrow). The septum is the posterior wal of the main chamber and extends to the crux cordis (*) AO=aorta

Fig. 6. (case-8) Univentricular heart of right ventricular type, absent left atrioventricular connection. The rudimentary chamber is a slit-like trabecular puch (TP) and located in the left posterior. The location of the rudimentary chamber and the site of the septum are well demarcated epicardially by the right and left delimiting arteries (RDA & LDA). In this case, since the posterior descending artery is the right delimiting artery, the ventricular loop is normally related (d-loop). LA=left atrium, LPV=left pulmonary vein, *=crux cordis

Fig. 7. (case-9) Univentricular heart of right ventricular type, absent right atrioventricular connection. The rudimentary chamber (RC) is a trabecular pouch with considerable space and fine trabeculae and located in the right posterior. Since the posterior descending artery is the left delimiting artery (LDA) of the rudimentary chamber, the ventricular loop is inverted (l-loop). AO=aorta, LA=left atrium, *=crux cordis

Fig. 8. Schematic representation of the disposition of the delimiting arteries; in univentricular heart of left ventricular type with normally related (A) and inverted (B) rudimentary chamber (RC); in univentricular heart of right ventricular type with normally related (C) and inverted (D) rudimentary chamber. AO=aorta, RA=right atrium, LA=left atrium, AD=anterior descending coronary artery, PD=posterior descending coronary artery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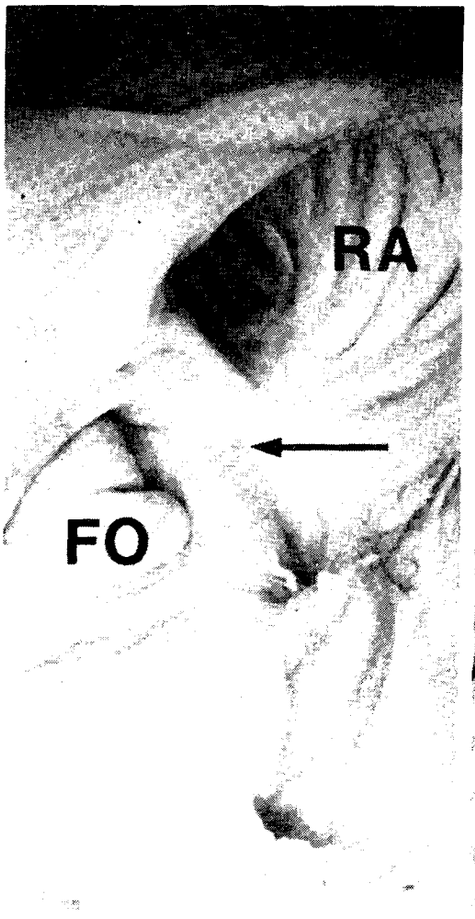
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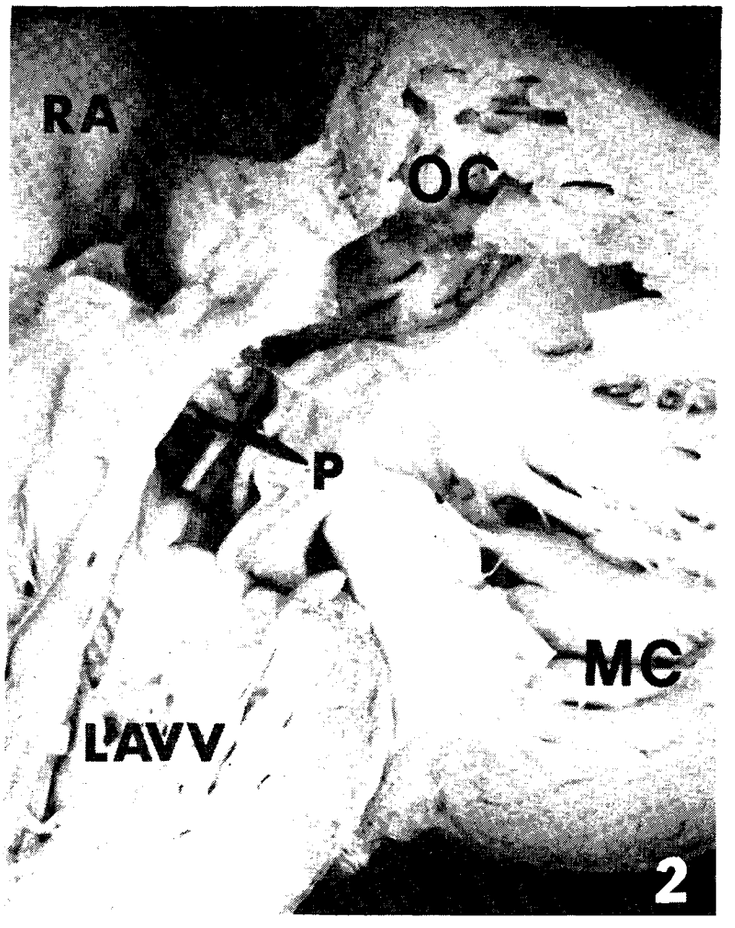
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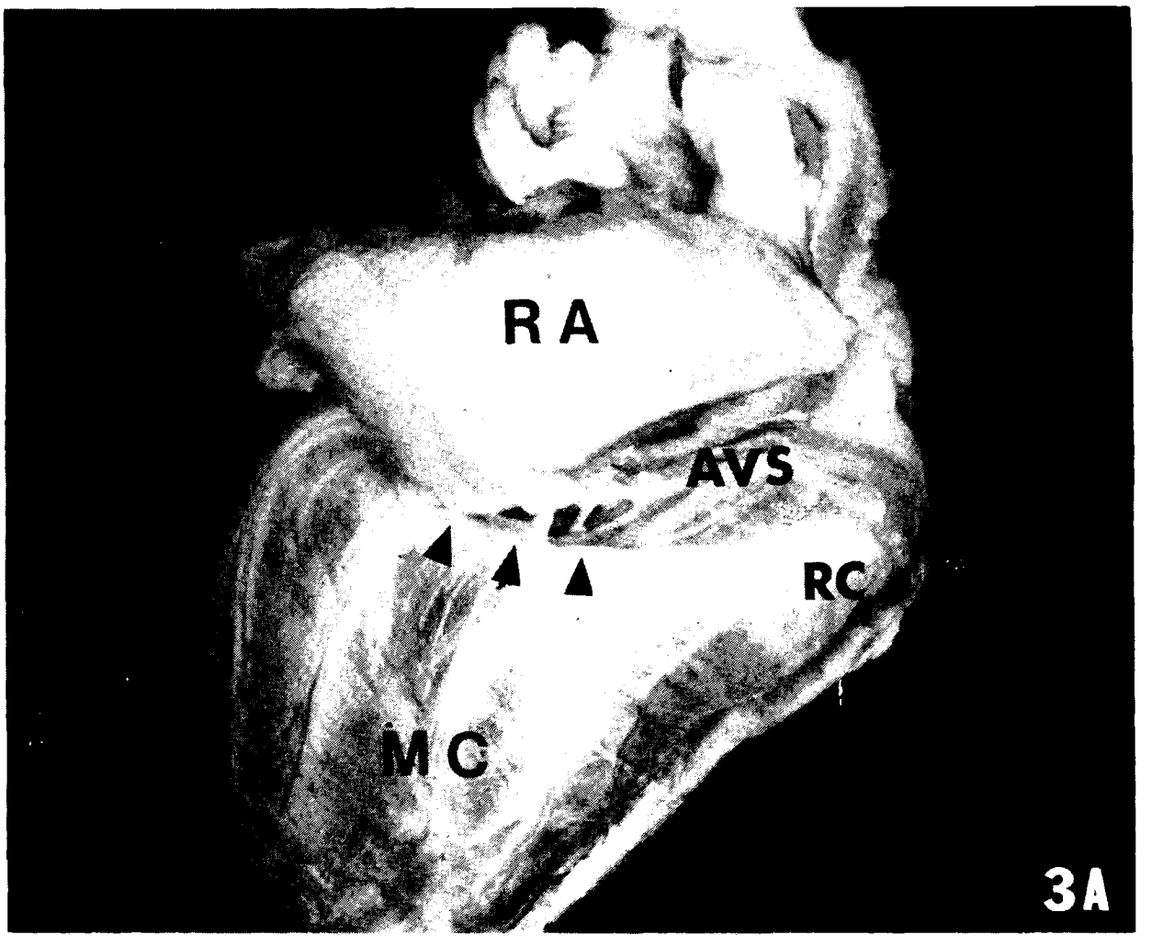
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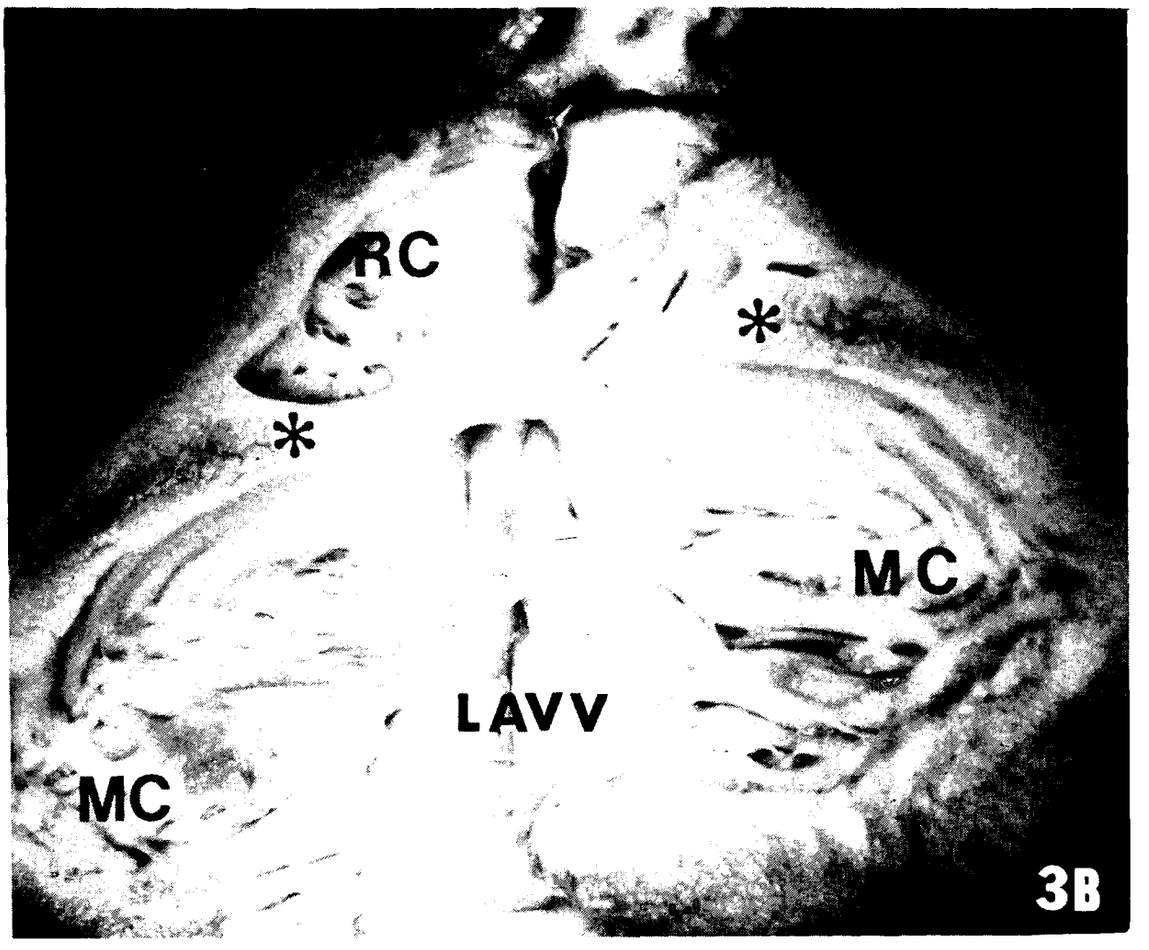
RC
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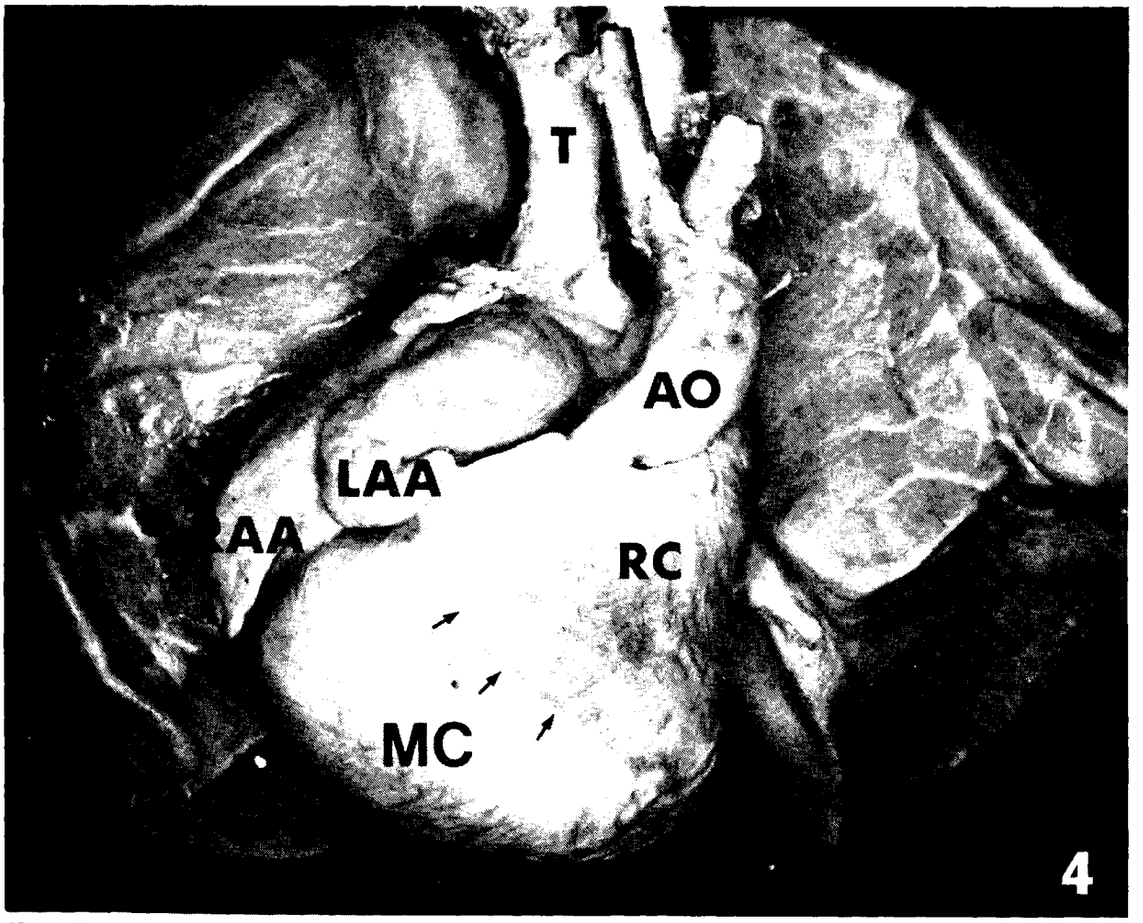
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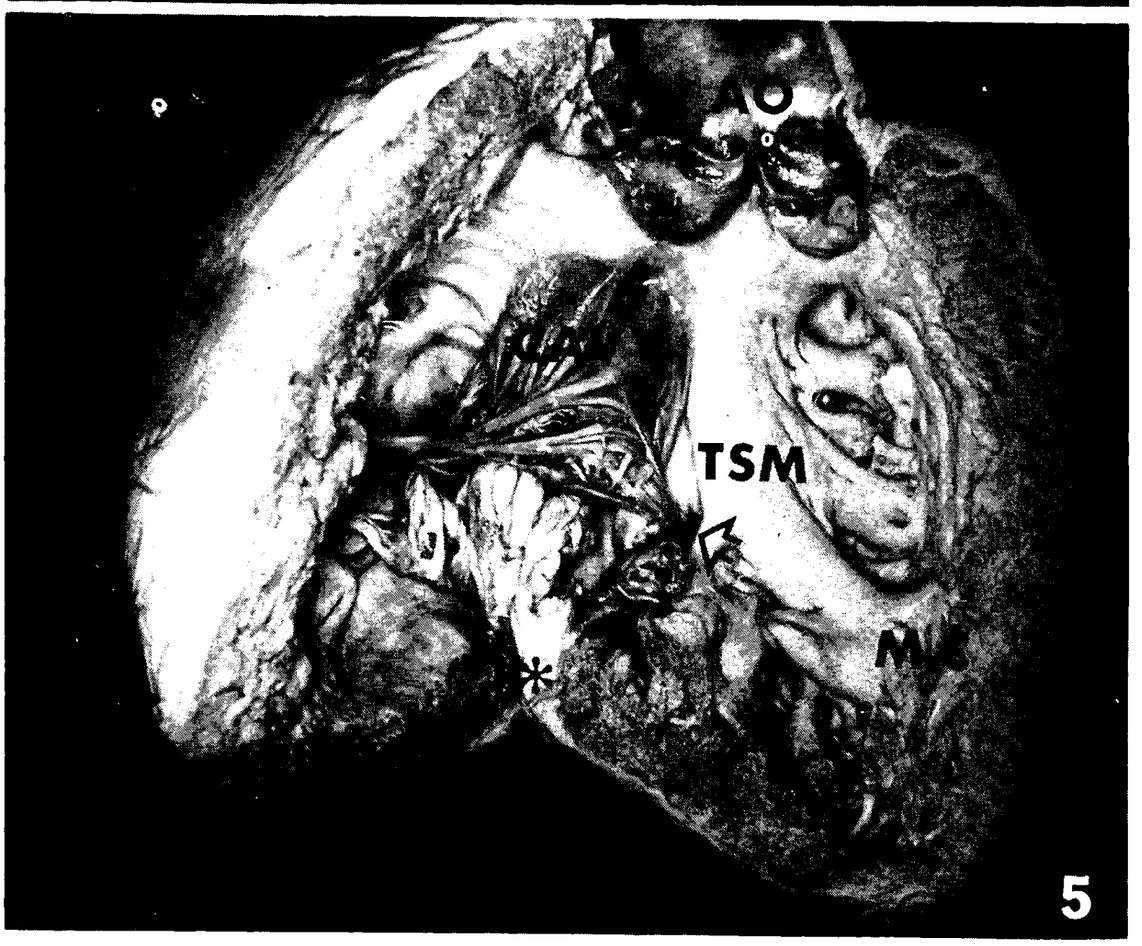
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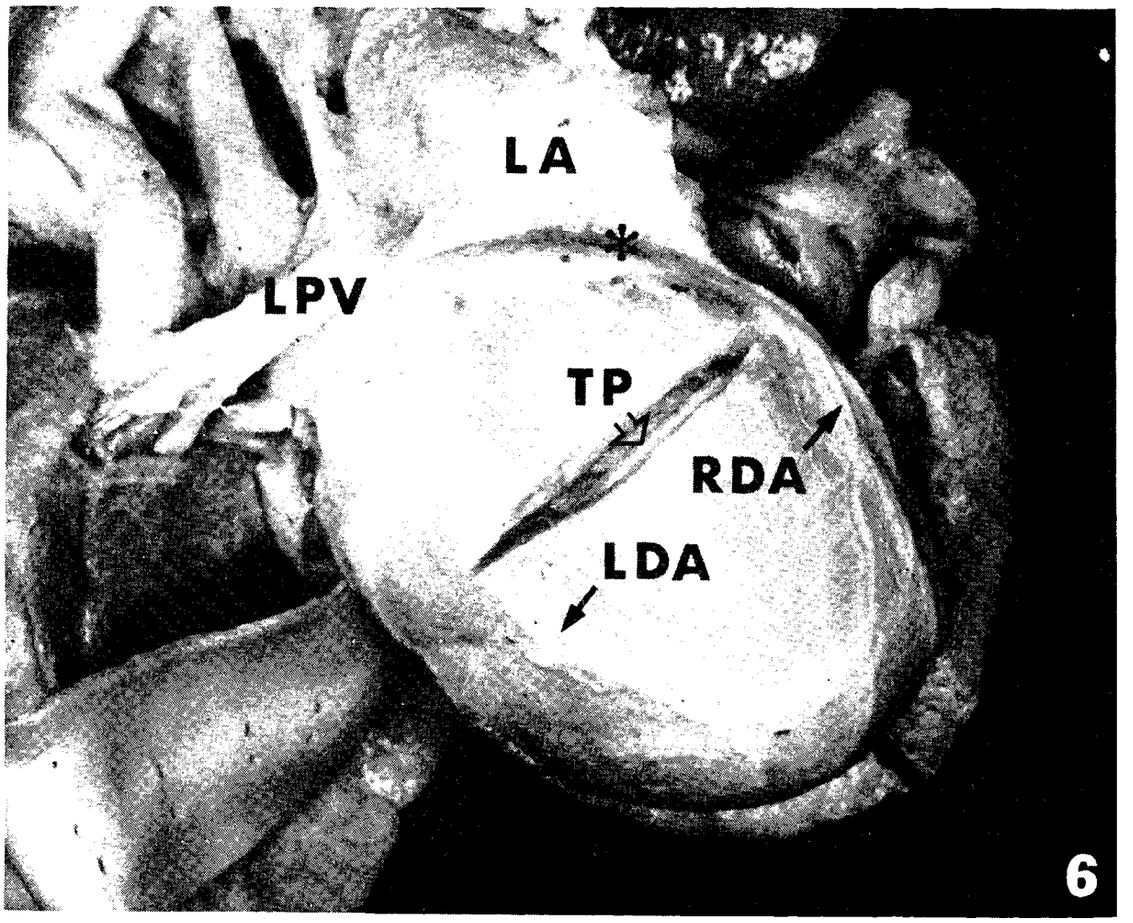
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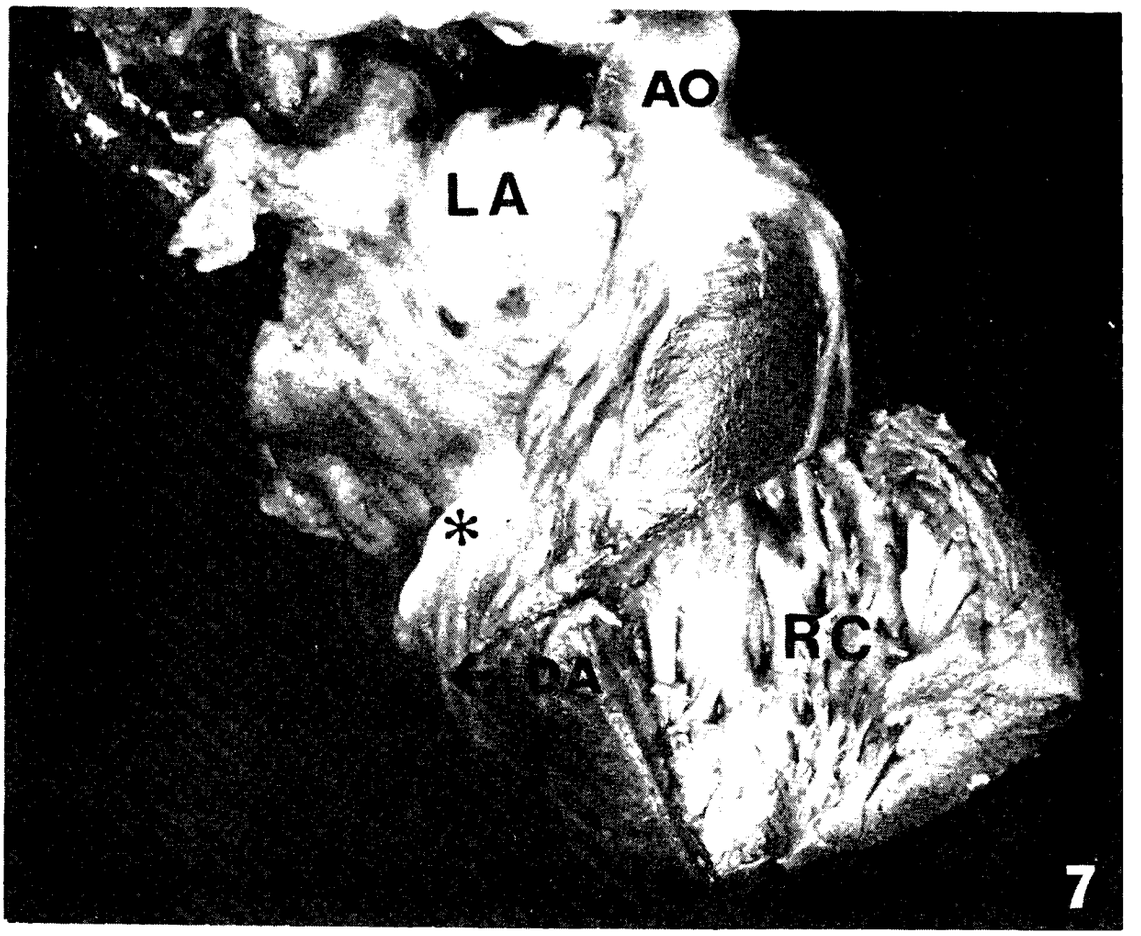
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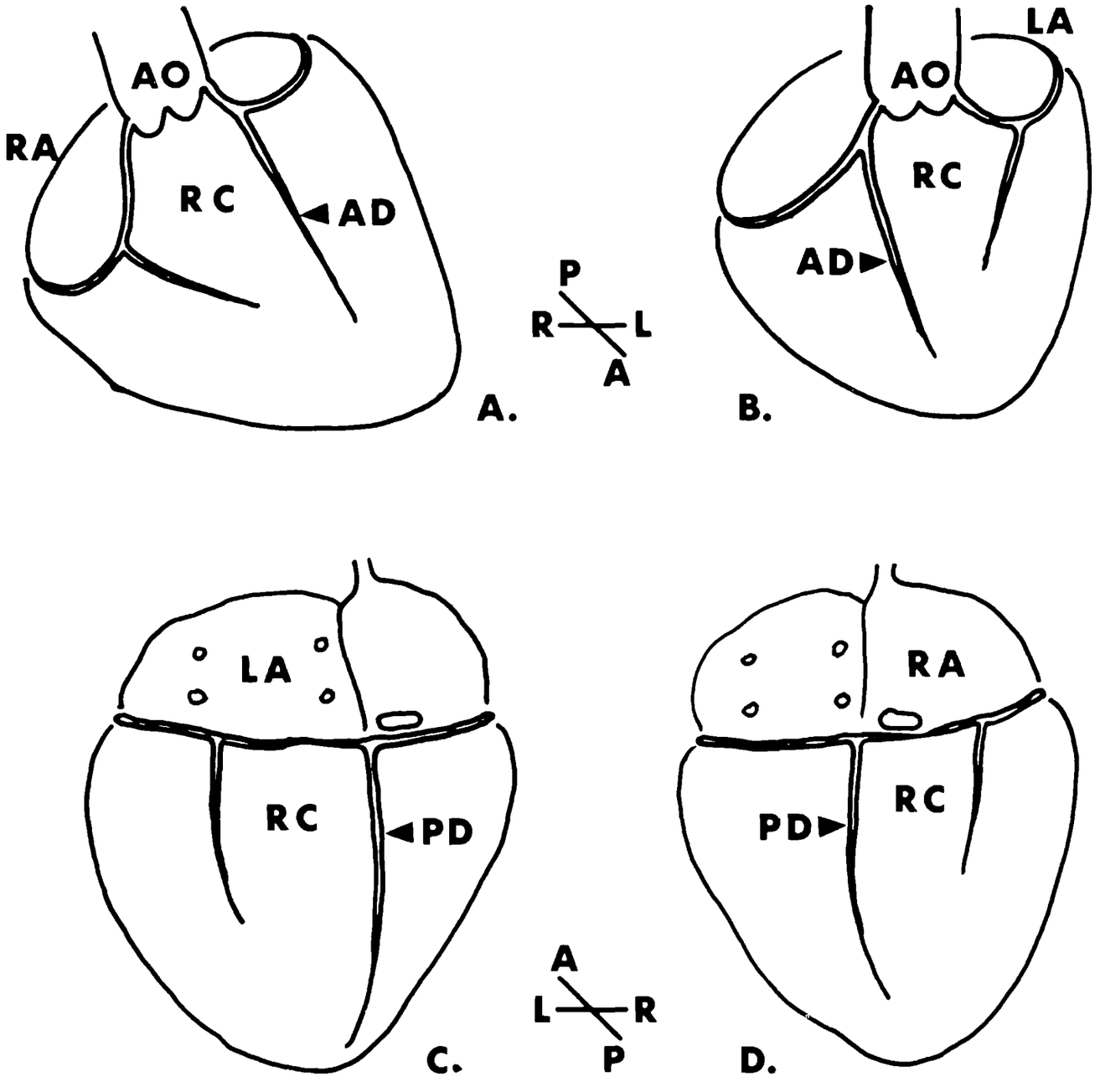
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