

# Anatomical variations in the coronary arteries.

## II. Less prevalent variations: Coronary anomalies

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### SUMMARY

With the introduction of new surgical procedures and the development of new techniques of cardiac imaging, the normal anatomy, variations and anomalies of coronary arteries have assumed new significance. The coronary arteries may present several variations, in terms of both number and position. The incidence of all coronary anomalies is 0.23% in autopsy series and ranges between 0.3% and 12% in angiographic series. Variations with a prevalence in the general population of less than 1% are therefore considered as coronary anomalies, and in many cases, their presence affects or may affect the subject's quality of life and even their survival. The coronary anomalies of the main coronary arteries are described following a criteria based on the origin of the anomalous vessel: ectopic aortic origin, origin from the pulmonary artery or origin from a systemic artery. Finally, the congenital coronary continuity and the coronary fistulas are reviewed.

**Key words:** Coronary arteries – Coronary anomalies

### INTRODUCTION

The coronary arteries may present several anomalies, in terms of both number and position. New image-based diagnostic techniques have led to greater reliability in the identification of

these anomalies, an in-depth knowledge of the normal anatomy of coronary arteries and their variations being required (Frommelt et al., 2001; McConnell et al., 1995; Post et al., 1995; Ropers et al., 2001).

The incidence of all coronary anomalies is 0.23% in autopsy series (Alexander and Griffith, 1956) and ranges between 0.3% and 12% in angiographic series (Chaitman et al., 1976; Engel et al., 1975; Barriales et al., 2001)., Owing to their relatively high rate of prevalence, some of these anomalies may be considered as variations within normal limits, taking the 1% presentation criteria as the limit between variations and anomalies (Levin, 1983; Angelini, 1989), and have already been described in the first part of this review paper (Reig, 2003).

Here, variations with a prevalence in the general population of less than 1%, and which are therefore considered as coronary anomalies, are described. In many cases, their presence affects or may affect the subject's quality of life and even their survival (Angelini et al., 1999; Basso et al., 2001).

### CORONARY ORIFICE ANOMALIES

#### *Atresia or hypoplasia of one or both coronary orifices*

The absence of one or both coronary orifices is usually associated with cases of pulmonary atresia (Lenox and Briner, 1972; Calder et al., 1987; Kasznica et al., 1987; Guenot et al., 1989), although cases of supra-avalvular aortic stenosis

have been described (Debich et al., 1989) and even as an isolated lesion (Saji et al., 1985). In the case of atresia or hypoplasia of an orifice, the blood reaches the non-atresic portion from the healthy coronary artery by collateral circulation, the functional result being equivalent to that of the single coronary artery. If both coronary orifices are affected, collateral circulation comes from extra-cardiac arteries, or directly from the cardiac cavities (Ueda et al., 1983).

#### *Single coronary orifice*

This anomaly, sometimes also known as single coronary artery, is characterised by the absence of the proximal portion of one of the coronary arteries, with the distal portion usually in its normal location (Ogden and Goodyer, 1970). Its existence was first noted by Columbus (1559), although Thebesius (1716) published the first description. The incidence of single coronary orifice not associated with congenital cardiopathies is low, and has been placed at 0.04% for the general population (Alexander and Griffith 1956) and 0.2% - 0.4% in angiographic series (Hillestad and Eie, 1971; Baltaxe and Wixson, 1977; Neufeld and Schneeweiss, 1983). A single coronary artery originating in the right aortic sinus is slightly more frequent than in the left (51% vs. 49%) (Ogden, 1968). The single right coronary artery also presents a higher number of pattern variations. An association between the presence of a single coronary orifice and anomalies in the aortic valve has been described, especially with the bicuspid (Hillestad and Eie, 1971) and tetracuspid (Kim et al., 1988) aortic valves.

From the single coronary orifice, four paths – preventricular, interarterial, transeptal and retro-aortic - may be followed by the anomalous artery (McAlpine, 1975). The least commonly described arterial path is the transeptal one (Figure 1), in which the anterior intraventricular artery, a branch of the single coronary artery originated in the right aortic sinus, crosses the *crista supraventricularis* and the interventricular septum. Descriptions of this variant have been published in a few cases (Bochdalek, 1867; Sanes, 1937; White and Edwards, 1948; Saner et al., 1984; Schulte et al., 1985; Reig et al., 1989; Dollar and Roberts, 1989). On this path, an increase in the elastification of the intimal layer of the transeptal vessel can be seen, together with a marked fibrosis of the middle layer, which gives it the appearance of a venous vessel (Reig et al., 1989).

The interarterial path of the single coronary artery has been associated with sudden death (Sharbaugh and White, 1974). In the other possible paths, its course is benign, although in the case of arteriosclerotic occlusion of the single artery, its consequences are often fatal (Roberts

1987, Kwok et al 2000). Both in the case of the interarterial path as well as the anterior path, there is also the possibility of lesion of the anomalous vessel during surgical manipulation of the pulmonary infundibulum, during valvular replacement, or during a coronary by-pass operation (Longenecker et al., 1961; Berry and McGoon, 1973; Kelley et al., 1977).

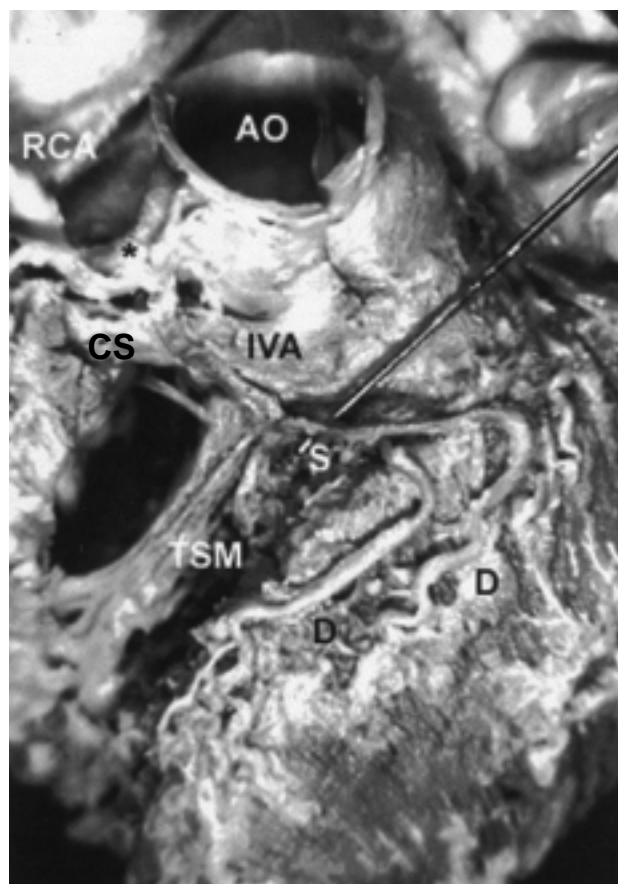
*Ectopic situation in a different aortic sinus.* – The specific anomalies of each artery are described below.

#### ANOMALIES OF THE LEFT CORONARY ARTERY

##### *Ectopic origin in a different aortic sinus.*

The left coronary artery originating from the right aortic sinus

This anomaly has been described as having an incidence of 0.02% in autopsy series, (Ale-



**Fig. 1.** Anterior surface of the heart after resection of the right outflow tract. A single coronary artery (\*) originates from the right coronary sinus. From the anomalous vessel, the anterior interventricular branch arises, crosses the *Crista Supraventricularis* and follows a transeptal path. On elevating the artery with a probe, several septal branches become visible, including the branch for the *trabecula septomarginalis*. (AO: aorta; CS: *crista supraventricularis*; D: diagonal or lateral branches; IVA: anterior interventricular branch; RCA: right coronary artery; S: septal branches; TSM: *trabecula septomarginalis*).

xander and Griffith, 1956), and between 0.05% and 0.19% in angiographic series (Chaitman et al., 1976; Kimbiris et al., 1978). As in the case with the location of both coronary orifices in the left aortic sinus, some authors consider this situation to be a variant of the single coronary artery, regardless of whether or not both arteries originate in a common orifice or not (Boucek et al., 1984).

The ectopic location of the left orifice in the right aortic sinus may present the following variations (Kragel and Roberts, 1988): (1) a common orifice with the right coronary artery, which is usually located above the aortic supra-ventricular ridge, and (2) an independent orifice, anterior or posterior to the right coronary artery origin.

The ectopic left orifice usually takes the form of a slit-like orifice in which the left coronary artery originates. This artery emerges forming a 180° angle with the aortic wall on the transversal plane (Figure 2). It may follow various paths from its origin (McAlpine, 1975; Ishikawa and Brandt, 1985; Greenberg et al., 1989). These are anterior, in front of the right ventricular infundibulum (Cheitlin et al., 1974; Liberthson et al., 1974), retroaortic (Murphy et al., 1987), interarterial, between the aorta and the pulmonary artery (Barth and Roberts, 1986), and finally, transeptal, through the *crista supra-ventricularis*



**Fig. 2.** The left coronary artery arises from an independent orifice located in the right aortic sinus and follows a retroaortic course. The aorta is displaced forwards and, as it arrives to the left coronary sinus the anomalous vessel bifurcates into the anterior interventricular and circumflexus branches. (AO: aorta; CX: circumflex branch; LCA: left coronary artery; RCA: right coronary artery; SVC: superior vena cava).

and the interventricular septum (Cheitlin et al., 1974; Roberts et al 1982).

In most of the cases described, the left coronary artery passes in front of the pulmonary infundibulum or between the aorta and the pulmonary artery, following an inter-arterial path (Kimbiris, 1985), the retroaortic path being less common (Figures 2 and 3). The inter-arterial path has been associated with sudden death, especially during or immediately after intense exercise (Cheitlin et al., 1974; Barth and Roberts, 1986). This is due to the compression of the ectopic orifice, or compression of the left coronary artery owing to the expansion of the aortic and pulmonary roots during intense exercise.

The anterior path of the large vessels does not present relevant haemodynamic problems. However, when this anomaly is associated with a tronco-conal congenital cardiopathy, such as Fallot's tetralogy, there is the possibility of accidental injury to the left coronary artery or the anterior interventricular branch during surgical handling of the pulmonary infundibulum.

The interarterial path of the ectopic left coronary artery rises slightly after its origin, followed by an anterior incurvation around the aortic root, between the latter and the pulmonary root. It then assumes a posterior concavity until it reaches the left coronary artery's normal position, where it divides into its final branches. Between the two great arteries, the left coronary artery is usually located below the level of the pulmonary valve (Liberthson et al., 1974).

The transeptal path is characterised by an intramyocardial route, almost from its starting point, crossing the *crista supra-ventricularis* and the upper part of the interventricular septum, and returns to an epicardial position at some point on the upper two thirds of the anterior interventricular sulcus. From this point onwards, it forks into its terminal branches – the anterior interventricular branch continues along the anterior interventricular sulcus, while the circumflex branch ascends along the anterior interventricular sulcus until its normal starting point, from where it continues on its normal path (Cheitlin et al., 1974; Roberts et al., 1982; Barth and Roberts, 1986; Reig et al., 1994). The transeptal path, on its intramyocardial route, typically follows an upper concavity, located below and behind the pulmonary valve (Ishikawa and Brandt, 1985). Often, on its intramyocardial route it supplies the first anterior septal artery, which has been considered as an angiographic sign of the interarterial and transeptal paths (Chaitman et al., 1976; Lipoff, 1988).

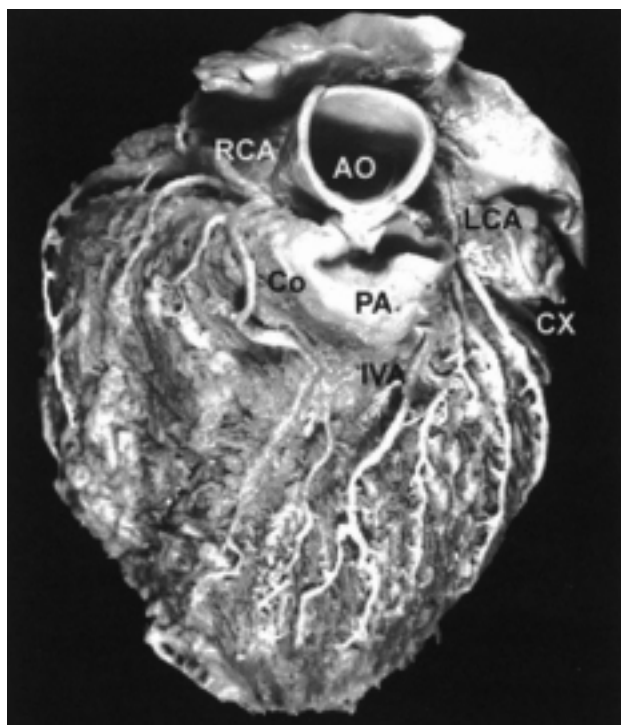
The retroaortic left coronary artery path of ectopic origin is the one least commonly observed. Its frequency of presentation, in terms of the total anomalous paths of the left coronary artery, is between 28% and 40% (Chaitman et al.,

1976; Kimbiris, 1985), although some authors refer a lower incidence (Ishikawa and Brandt, 1985; Click et al., 1989). In this path, the left coronary artery bends behind the aorta until it reaches its normal starting point, where it divides into its terminal branches (Figure 3). This anomaly has been observed associated with cases of persistent *truncus arteriosus comunis*, as well as a double outlet of the right ventricle (Vlodaver et al., 1975).

Ischemic complications have also been noted in cases of a retroaortic path of the left coronary artery, attributed to the angulation of the anomalous vessel towards the left, behind the aorta, and the systolic expansion of the aorta (Murphy et al., 1987). The possibility of injury during cardiac surgery to replace the aortic or mitral valve, when stitching a prosthesis, or as a result of compression of a mechanical prosthesis once surgery is over should also be considered (Roberts, 1987).

The left coronary artery originating in the posterior aortic sinus

The left coronary artery originating from the posterior aortic sinus is an extraordinarily rare anomaly, with very few descriptions of hearts with no other anomalies published (Dagonet,



**Fig. 3.** Same specimen as figure 2. Anterior surface of the heart showing the dissection of the coronary arterial tree. After a retroaortic course, the anomalous left coronary artery divides into the terminal branches: the anterior interventricular branch adopts its usual trajectory, but without reaching the *apex cordis*; the circumflex branch ends as the left marginal artery. (AO: aorta; Co: conal branch; CX: circumflex branch; IVA: anterior interventricular branch; LCA: left coronary artery; PA: pulmonary artery; RCA: right coronary artery).

1952; Ogden, 1968; Click et al., 1989; Ishikawa et al., 1990; Iñiguez Romo et al., 1991; Hamamichi et al., 2000). There are, however, some descriptions of cases associated with the transposition of the great arteries (Neufeld and Schneeweiss, 1983; Gittenberger et al., 1986).

*The left coronary artery originating from a systemic artery.*

Some cases of the left coronary artery originating from a systemic artery have been described, such as the ascending aorta (Di Guglielmo and Guttadauro, 1954) or the brachiocephalic trunk (Santucci et al., 2001). These are exceptional cases, most of them presenting an association with aortic arch anomalies. Most cases described of coronary origin in a systemic artery affect one of the left coronary artery's terminal branches (Robicsek et al., 1967).

*The left coronary artery originating from the pulmonary artery*

An anomalous coronary artery originating from the pulmonary artery was described for the first time by Brooks (1886) in the right coronary artery and by Abbott (1908) in the left coronary artery.

An anomalous coronary artery originating from the pulmonary artery is a relatively infrequent anomaly, with an incidence of 0.01% in the general population (Alexander and Griffith, 1956). The incidence of congenital cardiopathies in patients affected by cardiac heart disease has been placed at 0.5%, while among the population receiving angiographic exploration, it was 0.1% (Baltaxe and Wilson, 1977). The most commonly observed profile consists of the anomalous left coronary artery originating from the pulmonary artery, while the right coronary artery originates in the corresponding aortic sinus (Vlodaver et al., 1975). This combination is seen in 83% of cases of a anomalous coronary artery originating in the pulmonary artery (Boucek et al., 1984). In 1933, Bland, White and Garland described the clinical profile produced by this anomaly, coining a name for the syndrome. In this anomaly, the left coronary artery normally originates in the left posterior pulmonary sinus, which is the one facing the aorta. It is usually shorter in length than the cases of aortic origin, values of between 2 and 5 mm having been described. After this short path, the left coronary artery divides into its terminal branches, which follow their usual path and layout. The left coronary artery and its terminal branches are macroscopically similar in appearance to a vein, owing to their thin walls.

The anomalous origin of the left coronary artery from the pulmonary trunk together with the coronary fistulas are the congenital coronary anomalies that most commonly produce haemodynamic alterations.

Ectopic origins have also been described in the branches of the pulmonary arteries (Smith et al., 1989; Levin et al., 1990).

#### ANTERIOR INTERVENTRICULAR BRANCH ANOMALIES

##### *Anterior interventricular branch originating from the right aortic sinus*

The anterior interventricular branch may originate in an independent orifice in the right aortic sinus, usually in a forward position from the right coronary artery orifice, or less frequently, it may originate in the right coronary artery itself. The angiographic incidence of this anomaly varies between 0.03% and 0.02%, which represents 5% of all coronary arterial anomalies (Kimbiris et al., 1978; Click et al., 1989). The anterior interventricular branch originating in the right aortic sinus is usually observed in association with various congenital cardiopathies, such as ventricular inversion, transposition of the great arteries, double outlet of the right ventricle and is the most frequently observed abnormal arterial pattern in Fallot's tetralogy (Dabizzi et al., 1980; Greenberg et al., 1989; Neufeld and Schnee-weiss, 1983). Some cases have also been described in patients with no other anomaly (Roberts, 1987; Barriales et al., 2001).

The most frequently observed path of the anomalous vessel is the anterior one (Click et al., 1989), in which the anterior interventricular branch is situated in front of the infundibulum of the right ventricle, reaching the anterior interventricular sulcus and from that point following its normal path. The circumflex branch, with a normal path and location, originates in the orifice located in the left aortic sinus.

The anterior path of the anomalous vessel in the pulmonary infundibulum is very important to the surgeon since if it is not noticed, it may be cut if a surgical incision is made in the right ventricular infundibulum. Cases associated with congenital cardiopathies that require surgical remodelling of the right ventricular infundibulum are especially vulnerable. In a series of 27 cases of Fallot's Tetralogy, Berry and McGoan (1973) described 8 cases of sudden death during surgery as a result of myocardial infarction due to accidental cutting of the anomalous vessel. For this reason, both the exact location and the degree of twisting of the anomalous artery must be considered when choosing the surgical technique used to correct this congenital cardiopathy (Humes et al., 1987).

Cases have also been observed of an anomalous origin of the anterior interventricular branch with interarterial and transeptal paths (Roberts, 1987). These cases hold the same risks as those described when treating anomalous

paths of the left coronary artery, mainly in the interarterial path: the possibility of sudden death, especially during or immediately after intense exercise, either as a result of compression of the ectopic orifice or of the anterior interventricular branch itself (Cheitlin et al., 1974).

In the transeptal vessel, the anomalous vessel crosses the *crista supraventricularis* and the interventricular septum (Kimbiris et al., 1978; Virmani et al., 1989). This situation presents characteristics that are partially similar to those observed when the left coronary artery originates in the right aortic sinus, or in cases of a single orifice located in the right aortic sinus, both with a transeptal path of the main left trunk. The transeptal path of the anterior interventricular artery of ectopic origin is a rarely observed anomaly, and only a few cases have been described (Bochdalek, 1867; Sanes, 1937; White and Edwards, 1948; Saner et al., 1984; Schulte et al., 1985; Reig et al., 1989; Virmani et al., 1989; Dollar and Roberts, 1989), as well as some angiographically diagnosed cases (Kimbiris et al., 1978). Structural changes, consisting of a thickening and elastification of the intimal layer of the arterial wall, together with a marked fibrosis of the middle layer in the intramyocardial portion of the anomalous vessel have been observed (Saner et al., 1984; Schulte et al., 1985; Reig et al., 1989). These are thought to be the arterial wall's response to the mechanical overload to which it is subjected by the myocardial fibers. These structural alterations afford the intramyocardial portion a macroscopic appearance similar to a vein.

The anterior interventricular branch, having originated in an independent orifice, in the same orifice as the right coronary artery or as a branch of it, follows a similar path in all the cases described. The artery undergoes a 90° forward angle, penetrating the myocardium and crossing the *crista supraventricularis* in an oblique direction downwards and to the left, until it reaches the same level as the upper septal papillary muscle, where it changes direction, following a horizontal path. Then, it crosses the upper part of the interventricular septum and returns to an epicardial position in the anterior interventricular sulcus, where several lateral branches (diagonal arteries) begin. Along its intramyocardial path, it supplies several septal arteries, especially those responsible for irrigating the right branch of the atrio-ventricular bundle, the *trabecula septomarginalis*, and the anterior papillary muscle of the right ventricle. In most cases described, the circumflex branch originates in the same orifice as the right coronary artery, or as a branch of it, and follows a retroaortic path until it reaches the left coronary sulcus

### *The anterior interventricular branch originating from a systemic artery*

Some cases of the anterior interventricular branch originating from a systemic artery have been described, such as the common carotid artery, the subclavian artery or the internal thoracic artery (Evans, 1933; Robicksec, 1967). These cases were associated with aortic arch anomalies or with tronco-conal cardiopathies, such as the Tetralogy of Fallot. From the anomalous origin, the vessel perforates the pericardium and reaches the anterior interventricular sulcus. Usually, the rest of coronary arteries also show some anomalies of origin or trajectory.

### *The anterior interventricular branch originating from the pulmonary artery*

The anterior interventricular branch originating from the pulmonary artery is a rarely observed anomaly and is a variant of the Bland-White-Garland syndrome (Schwartz and Robisek, 1971; Probst et al., 1976; Baltaxe and Wixson, 1977; Donaldson et al., 1979; Singh and Taylor, 1983; Evans and Phillips, 1984; Tamer et al., 1984; Roberts and Robinowitz, 1984; Reig and Ruiz de Miguel, 1990). In the published series, the anterior interventricular branch originates in the left posterior pulmonary sinus, after which the anomalous vessel is located in the correspondent sulcus, and follows its usual path.

The circumflex branch may vary in origin – in the left aortic sinus through a supernumerary orifice, or in the right coronary artery (Schwartz and Robisek, 1971; Donaldson et al., 1979; Reig and Ruiz de Miguel, 1990). In the latter two cases, the circumflex branch follows a retroaortic path until it reaches until it reaches the left coronary sulcus.

## CIRCUMFLEX BRANCH ANOMALIES

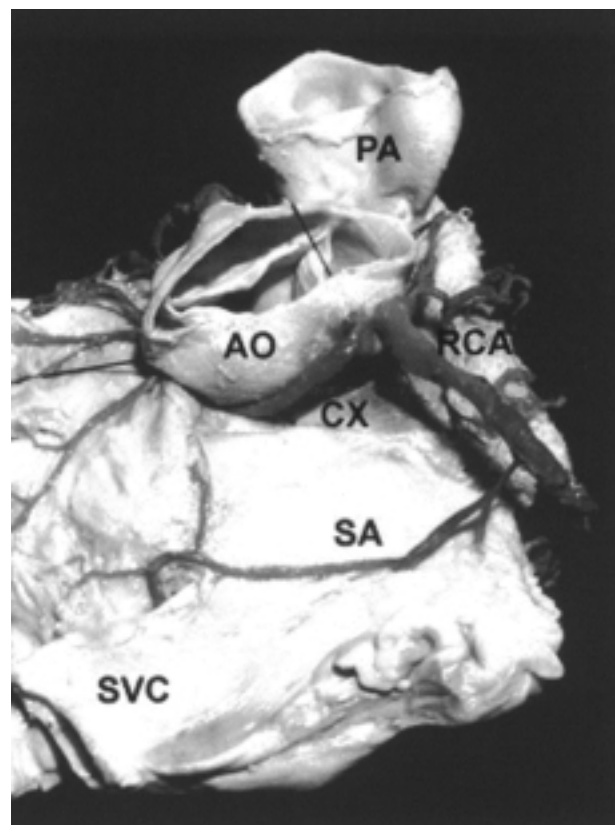
### *The circumflex branch originating from the right aortic sinus*

The anomalous origin of the circumflex branch from the right aortic sinus is the most prevalent congenital coronary anomaly. An incidence varying between 0.45% and 0.70 has been described in angiographic series (Chaitman et al., 1976; Vlodaver et al., 1975; Engel et al., 1975; Click et al., 1989; Iñiguez Romo et al., 1991). However, some authors, such as Ogden (1968), Page et al. (1974), Roy et al. (1975), and Gensini (1975) consider that the incidence among the general population is greater than that described, reaching 1%, and that it should therefore be considered as an anatomical variation rather than an anomaly. As regards its association with the various congenital cardiopathies, it has been described as the second most common pattern of coronary anomaly associated with transposition

of the great arteries (Neufeld and Schneeweiss, 1983), having also been observed in cases of double outlet right ventricle and persistent *truncus arteriosus communis* (Vlodaver et al., 1975).

Antopol and Kugel, in 1933 published the first description of the ectopic origin of the circumflex branch. Later, Page et al. (1974) described three circumflex branch variations of ectopic origin in the right aortic sinus. These were (1) originating in an independent orifice located in the right aortic sinus and generally behind the right coronary artery orifice (Figure 4), (2) originating in a common orifice with the right coronary artery, and (3) originating as a collateral branch of the right coronary artery (Figure 6). The frequency of each of these varieties is similar, although from the analysis of the various cases published (Alexander and Griffith, 1956; Page et al., 1974; Kimbiris et al., 1978) a mild predominance of the latter two can be seen.

The clinical significance of this anomaly is a matter of controversy. While some authors say that this anomaly has no pathological significance *per se*, (Neufeld and Scheneeweiss, 1983; Roberts, 1987; Molajo et al., 1988), other authors consider that it is an anomaly that may lead to an alteration in cardiac perfusion (Gallet et al., 1986; Silverman et al., 1978; Piovesana et al., 1989; Samarendra et al., 2001).



**Fig. 4.** Anomalous origin of the circumflex branch from the right aortic sinus. (AO: aorta; CX: circumflex branch; PA: pulmonary artery; RCA: right coronary artery; SA: sinuatrial nodal branch; SVC: superior vena cava. Image provided by Dr. J.R. Sañudo).

Regardless of its possible role as a producer of ischemic myocardia, the lack of recognition of this anomaly may have clinical consequences, when failure to see the contrast in circumflex branch on a coronary angiography, when this vessel originates in an independent orifice, is interpreted as an obstruction of this branch (Page et al., 1974; Engel et al., 1975). Given the relative frequency of this anomaly, when the circumflex branch cannot be seen in its normal position during coronary angiography, the possibility of an ectopic origin should always be taken into consideration.

Moreover, the possibility of injury to the circumflex branch during cardiac surgery to replace the aortic or mitral valves, when stitching prostheses or as a result of compression of a mechanical prostheses once surgery is over should also be considered (Roberts and Morrow, 1969). Recognition of this anomaly is also vital for correct coronary perfusion during cardiac surgery (Lillehei et al., 1964; Page et al., 1974; Roberts, 1987).

In any of the three variations, the path of the circumflex branch is always the same – after its origin, it immediately goes backwards and to the left, around and behind the aorta, and is initially situated between the aortic posterior wall and the anterior wall of the right atrium, and subsequently the left atrium, until it is located in the left portion of the coronary sulcus, covered by the left auricular appendix, and finally adopts its usual path (Figures 5, 6). On this trajectory, the circumflex branch is related to the tricuspid and mitral rings, with the artery located in their septal portions, and also to the posterior aortic sinus, which is located in front of the anomalous artery.

The size and area of distribution of the anomalous circumflex branch are variable, as in the case of the circumflex branch originating in the left coronary artery (Page et al., 1974; Sañudo et al., 1989), and are related to the distribution areas of the anterior intraventricular branch and the right coronary artery.

#### *The circumflex branch originating from the pulmonary artery*

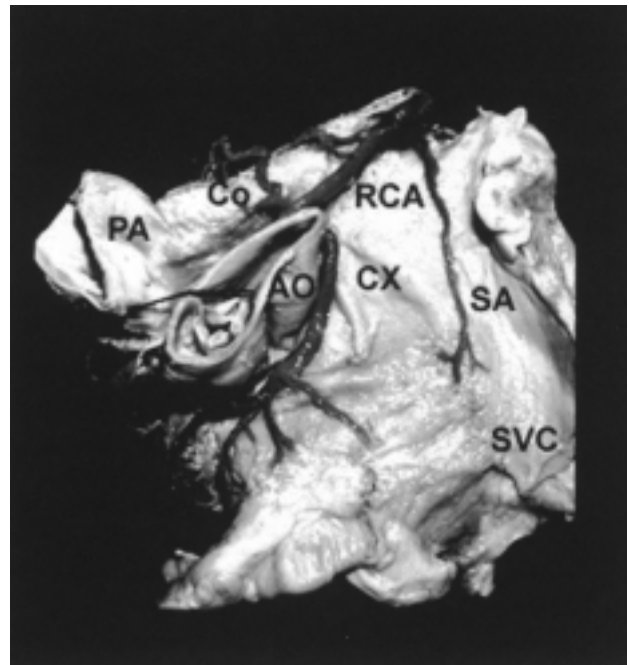
There are very few cases of anomalies of the circumflex branch originating from the pulmonary artery, with most of them found in infancy or adolescence (Roberts, 1987; Krishnamoorthy and Rao, 2001).

## RIGHT CORONARY ARTERY ANOMALIES

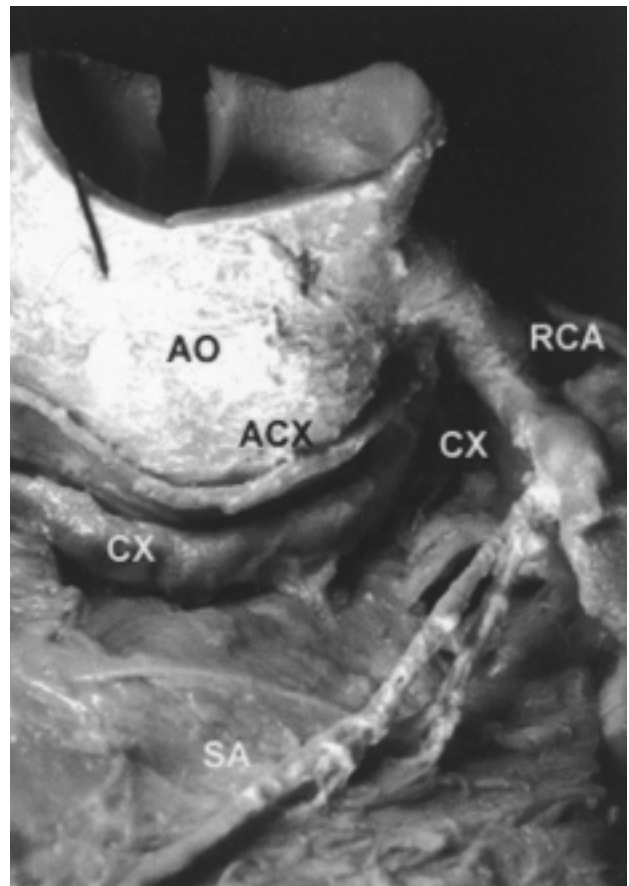
### *Ectopic origin in a different aortic sinus*

The right coronary artery originating from the left aortic sinus

This anomaly has been observed with an incidence of 0.17% - 0.38% in angiographic series



**Fig. 5.** The path of the anomalous circumflex branch, around and behind the aorta, situated between the posterior wall of the aorta and the anterior wall of the right atrium. (AO: aorta; Co: conal branch; CX: circumflex branch; PA: pulmonary artery; RCA: right coronary artery; SA: sinuatrial nodal branch; SVC: superior vena cava. Image provided by Dr. J.R. Sañudo).



**Fig. 6.** Origin of the circumflex branch from the right coronary artery. The anomalous vessel adopts a retroaortic course and gives off an atrial circumflex branch, with a parallel course (ACX: atrial circumflex branch; AO: aorta; CX: circumflex branch; RCA: right coronary artery; SA: sinuatrial nodal branch). x 2.5.

(Chaitman et al., 1976; Kimbiris et al., 1978; Topaz et al., 1992), and with an incidence of 0.03% in a necropsy series (Alexander and Griffith, 1956). The incidence in necropsy series is somewhat greater than that of the left coronary artery originating in the right aortic sinus although its angiographic incidence is lower. This is probably due to the fact that patients affected by the former anomaly usually present ischemic complications more frequently.

Some authors consider this situation to be a variety of the single coronary artery, regardless of whether or not both arteries originate in a common orifice (Boucek et al., 1984). The ectopic location of the right orifice in the left aortic sinus presents some variants (Kragel and Roberts, 1988). These are:

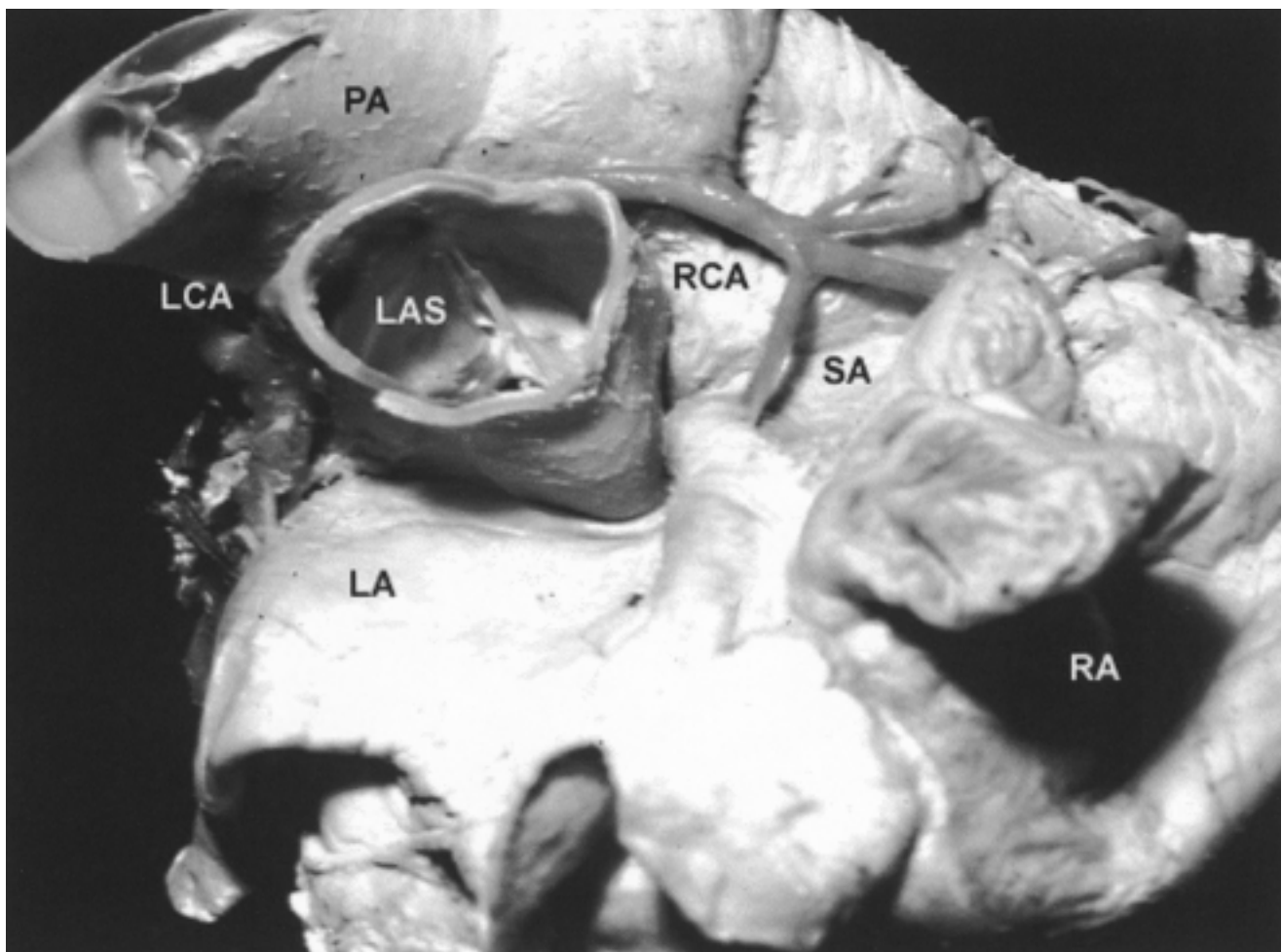
Originating from an independent orifice next to the left coronary one, usually in a more anterior position near the commissure between the left and right aortic valves (Figure 7). In this position, the orifice may be found at the same level as the supravulvar ridge, or above or below it.

Originating halfway between the left and right aortic sinuses, above the valvular commissure that joins them.

Originating in a common orifice with the left coronary artery, which may be located in the left aortic sinus, or halfway between the left and right aortic sinuses.

The ectopic right orifice usually presents a larger vertical diameter, from where the right coronary artery originates, which is usually located between the pulmonary artery and the aorta, following an interarterial path (Figure 8). From its beginnings, the ectopic right coronary artery undergoes a marked angulation, forming an acute angle with the aortic wall on the transversal plane (Roberts, 1987), and then goes towards the right. It is usually located between the aorta and the pulmonary artery, following a path towards the right coronary sulcus (Kimbiris et al., 1978; Neufeld and Schneeweiss, 1983; Sañudo et al., 1998). Some cases in which the right coronary artery's path lead to it being located in front of the pulmonary artery (Click et al., 1989) or behind the aorta (Neufeld and Schneeweiss, 1983) have also been described.

This anomaly was considered relatively benign in the absence of arteriosclerosis (Chaitman et al., 1976) until Benge et al., in 1980, warned of the possibility that it may cause myocar-



**Fig. 7.** Upper view of the heart showing both coronary orifices located in the left aortic sinus and the initial portions of left and right coronary arteries. (LA: left atrium; LAS: left aortic sinus; LCA: left coronary artery; PA: pulmonary artery; RA: right atrium; RCA: right coronary artery; SA: sinu-atrial nodal branch). x 1.5.



dic ischemia as a result of the artery's angulation after its origin, which in the case of a large aortic expansion, such as that observed during intense exercise, would lead to a compression of the vessel, giving rise to a mechanism that would end up occluding the ectopic orifice (Cheitlin et al., 1984; Virmani et al., 1986).

The right coronary artery originating from the posterior aortic sinus

The right coronary artery originating from the posterior aortic sinus is the most commonly found coronary distribution in cases of transposition of the great arteries (Neufeld and Schneeweiss, 1983; Gittenberger-de Groot et al., 1986), but it is a rarely observed anomaly in hearts with no other alteration (Vlodaver et al., 1975; Mahowald et al., 1986). Analysis of published cases does not show that this ectopic origin may have any pathological consequence.

*The right coronary artery originating from another coronary artery*

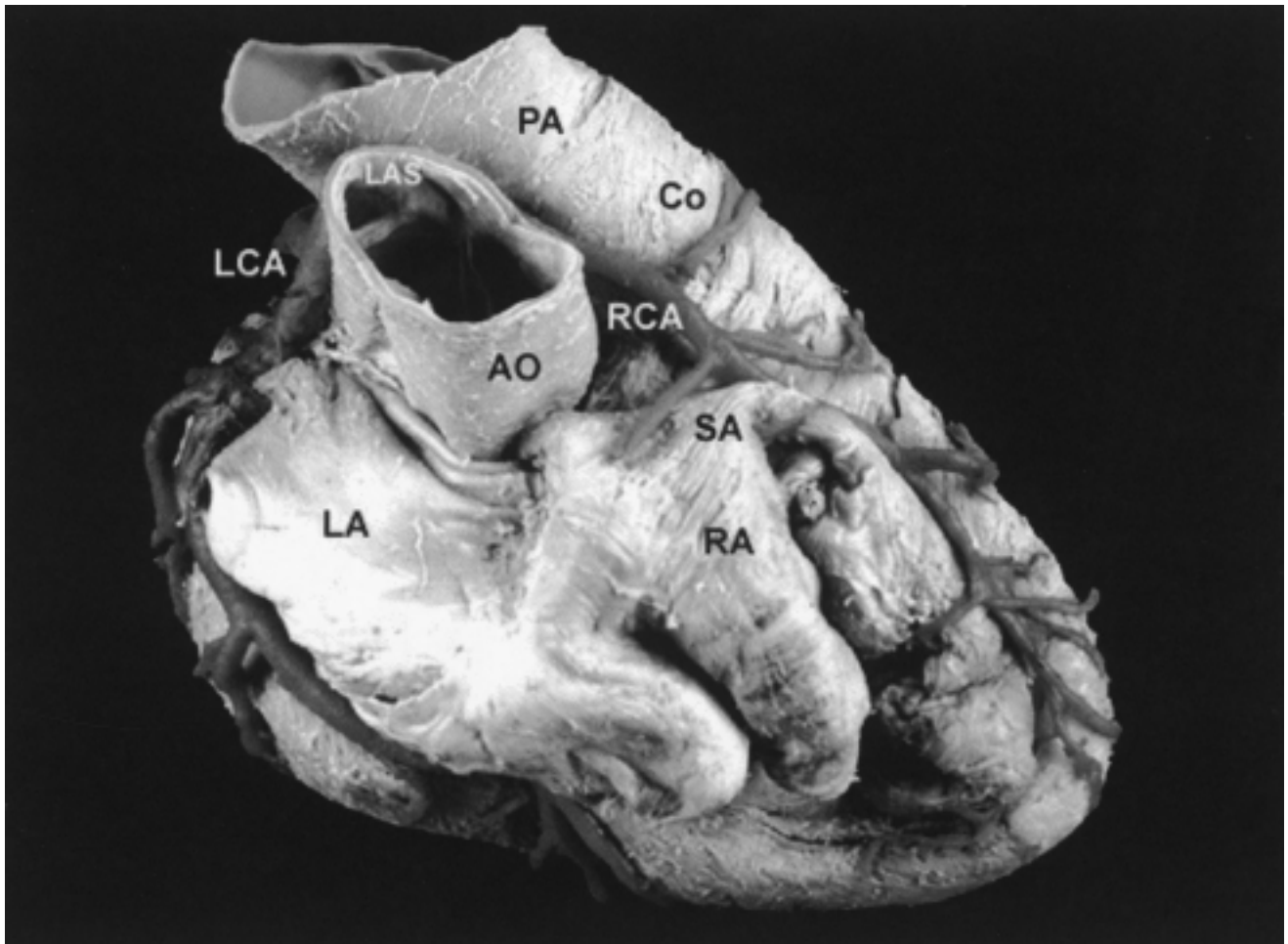
This is a group of very infrequent anomalies, generally found during angiographic exploration. The right coronary artery originating from

the left coronary artery, a few millimetres from its beginning, has been described (Barbour and Roberts, 1985). This location is in fact similar to that of the right coronary artery originating in the left aortic sinus, and strictly speaking, is a case of a single coronary artery with a left-sided origin.

The right coronary artery may also originate from the anterior interventricular branch (Simkoff et al., 1982; Amsel and Van der Mast, 1986; Habbab et al., 1987) or from an anterior septal branch (Meyers et al., 1984). In these cases, the artery's path passes in front of the pulmonary infundibulum. It may also originate from the circumflex branch or as a continuation of it (Tavernarakis et al., 1986; Sheth et al., 1988).

*The right coronary artery originating from a systemic artery*

There are very few of these cases described. Among them are origins in the ascending aorta, more than 2 cm from the corresponding sinus (Goldstein et al., 1990) or from another aortic sinus (Miller et al., 1987) or originating in the thoracic aorta (Cheatham et al., 1987).



**Fig. 8.** Ectopic origin of the right coronary artery arising from the left aortic sinus. The anomalous vessel follows an interarterial path, between the aorta and the pulmonary artery. (AO: aorta; Co: conal branch; LA: left atrium; LAS: left aortic sinus; LCA: left coronary artery; PA: pulmonary artery; RA: right atrium; RCA: right coronary artery; SA: sino-atrial nodal branch).

*The right coronary artery originating from the pulmonary artery*

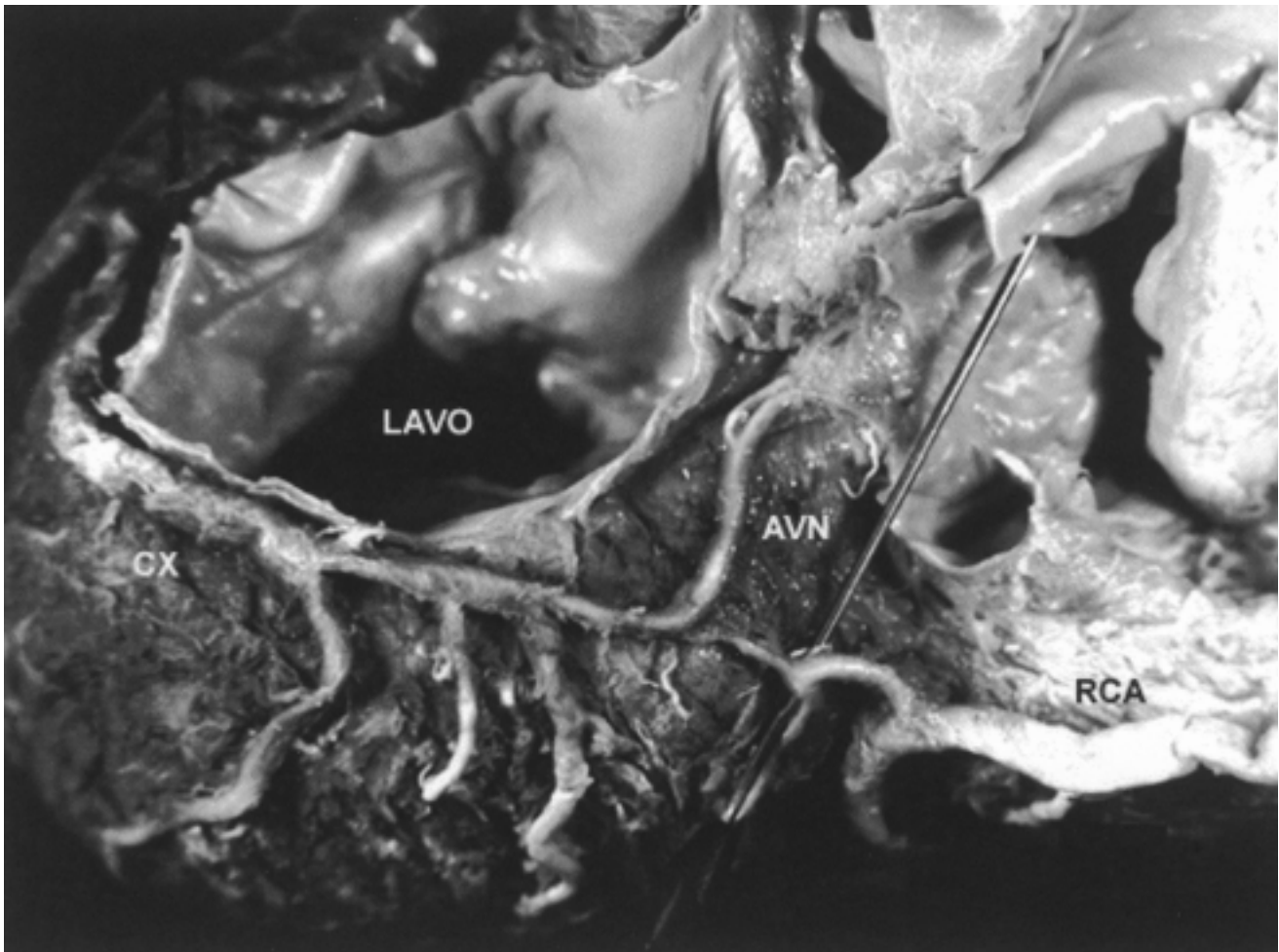
The first description of the right coronary artery originating from the pulmonary artery was given by Brooks in 1886. It is much less common than an anomalous origin of the left coronary artery, and accounts for between 7% and 9% of cases of coronary arteries of anomalous origin (Ogden, 1968; Boucek et al., 1984). In most published cases the orifice is located high in the right pulmonary sinus, above the supra-ventricular ridge. From its ectopic origin, its path and layout are normal. Its vascular wall is thin, similar to that of a vein, and usually has a dilated and twisted appearance. The left coronary artery and its branches also have an increased volume.

An anomalous origin of the conus branch from the right pulmonary sinus has also been described, with the same layout as that of the artery originating in the aorta. It is usually connected with other coronary arteries with a normal origin and acts as a fistulous communication between the high pressure coronary system and the low pressure pulmonary system (Ogden, 1968).

## INTERCORONARY CONTINUITY

Intercoronary continuity is not a common condition, and thus may be considered to be an anomaly, which gives the coronary circulation an "open end" character, with connections between two or more main arterial trunks, as is the case of cerebral circulation, or in the extremities, when under normal conditions, coronary artery circulation is "closed end", without a good established connection between the two main coronary arteries (Esente et al., 1983). It is found in subjects with no obstructive coronary lesions, although it may occasionally be found accompanying congenital or acquired coronary pathologies (Donaldson and Isner, 1984), when only the conduction, extra-mural and epicardial vessels are involved.

In most cases, intercoronary continuity is probably congenital and its path, which is the continuation of the arteries it connects, generally follows a straight line. Only two vessels are involved, which may or may not be from the same coronary artery. Functional factors encouraging the condition should not be ruled out.



**Fig. 9.** The atria have been removed in order to improve the visualization of the posterior coronary sulcus. A probe is placed under the communicating vessel (coronary arch) between the right coronary artery and the circumflex branch. (AVN: atrioventricular nodal branch; CX: circumflex branch; LAVO: left atrioventricular orifice; RCA: right coronary artery). x 1.5.

Basically, two locations have been described for intercoronary continuity – a communication between the circumflex branch and the right coronary artery, and a communication between the two interventricular branches.

#### *The coronary arch*

This is the name used for the communication between the right coronary artery and the circumflex branch, by means of a single anastomotic channel, providing continuity between these arteries (Greenberg et al., 1989). It is the intercoronary continuity location that is most frequently described (Figure 9). It was mentioned for the first time by Ruyschii (1701-1715).

Only a few cases from coronary angiography (Hines et al., 1981; Kutcher et al., 1982; Phillips and Berman, 1984; Voci et al., 1987) or from dissection (Reig et al., 1995) have been described, most of which have no signs of ischemic cardiopathy as their common feature.

#### *Continuity between both interventricular branches*

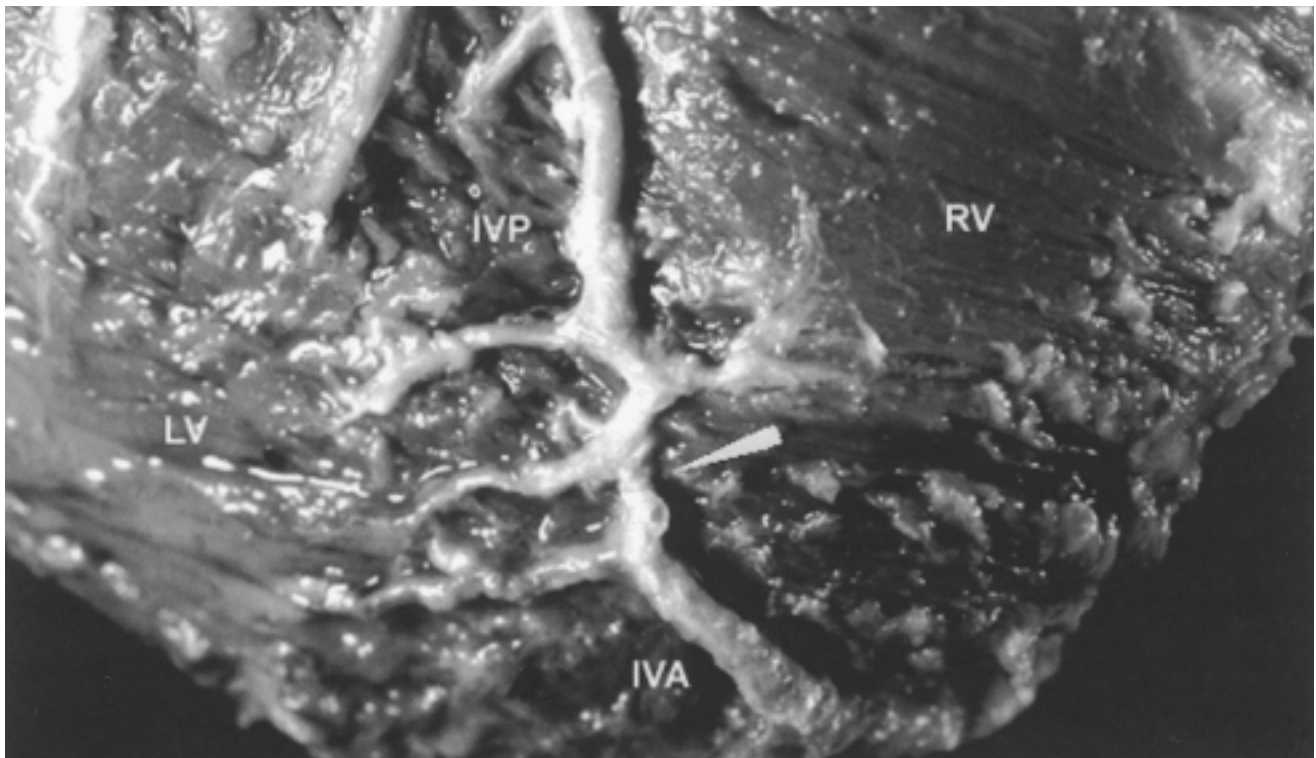
The connection between the anterior interventricular branch and the posterior interventricular branch by means of a straight anastomotic vessel that is always over 1.0 mm in diameter throughout the connection has been described in the distal portion of the posterior interventricular sulcus (Figure 10) (Linsenmeyer and Schneider, 1983; Donaldson and Isner, 1984; Reig et al., 1995).

In the absence of occlusive arterial lesions, the difference between intercoronary continuity

and the presence of arterial anastomosis can be clearly established by studying the vessel's morphology. In the case of anastomotic connections, the vessel looks like an arteriole, with an endothelium sustained by muscular and elastic collagenous fibres with little organisation. However, in the case of intercoronary continuity, the vessel presents the typical well-organised layers of a muscular-type artery (Donaldson and Isner; 1984). In cases where intercoronary continuity is seen in hearts affected by occlusive arterial pathology, it does not seem probable, knowing the histological structure in non-pathological hearts, that intercoronary continuity is the result of a transformation of pre-formed channels as a result of ischemic stimuli. The work of Schaper et al. (1988) show that even when the transformation of these channels takes place in vessels with a macroscopic arterial appearance, they cannot be considered as structurally normal arteries from the histological point of view. Moreover, their rectilinear appearance is very different from the tortuous collaterals that are found at epicardic level (Hines et al., 1981).

#### TERMINATION ANOMALIES – CONGENITAL CORONARY ARTERIAL FISTULAS

A communication between a coronary artery and a cardiac cavity or any part of the pulmonary or systemic circulation is known as a coronary fis-



**Fig. 10.** Diaphragmatic surface of the heart. The figure shows the continuity (arrowhead) between the anterior and posterior interventricular branches, located in the distal portion of the posterior interventricular sulcus. (IVA: anterior interventricular branch; IVP: posterior interventricular branch; LV: left ventricle; RV: right ventricle). x 3.5.

tula (Levin, 1983; Boucek et al., 1984), although use of this name is currently reserved for communications where there is an increase in the diameter of the vessel receiving the fistulous communication or where signs of volumetric overload of the affected cavity can be observed. This definition excludes arterio-sinusoidal communications with a small diameter at arteriolar level, as well as the thebesian veins, which are particularly common in the right cardiac cavities (Angelini et al., 1999).

The incidence of coronary fistulas in angiographic series varies between 0.1% and 0.2% (Said et al., 1997; Barriales et al., 2001). They account for 0.40% of congenital cardiopathies (Neufeld and Schneeweiss, 1983). Fistulous communications may originate both in the right and left coronary arteries, although a slightly higher incidence has been described in the left artery. The vessel of origin is dilated and twisted in appearance, owing to the increase in blood flow. The termination point of arterial fistulas is located mostly in the right cavities or in the pulmonary artery (Arani et al., 1978; Levin, 1983; Bosc et al., 1985; Said et al., 1997).

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