Congenital Anomalies Involving the Coronary Sinus

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Within the considerable body of information pertaining to congenital cardiac malformations, anomalies involving the coronary sinus have received relatively little attention. Although not always patent or functionally significant, some of these anomalies may assume considerable importance. They may occur as isolated and benign conditions or as a component of a variety of significant malformations. Failure to recognize anomalies in which the coronary sinus is involved may give rise to serious misinterpretation of cardiac catheterization data, and the associated altered hemodynamics may cause troublesome effects during surgical procedures.

This communication is intended to be a comprehensive study of the recognized anomalies of the coronary sinus. It is based largely on an analysis of necropsy specimens from the Cardiovascular Registry of The Charles T. Miller Hospital and the Department of Pathology of the University of Minnesota. A few conditions, of which we had no examples, are taken from the literature in order to complete this comprehensive report.

A classification of anomalies of the coronary sinus is presented in table 1 and illustrations of each anomaly will be presented in diagrammatic form.

Enlargement of the Coronary Sinus

Enlargement of the coronary sinus may occur in situations not necessarily anomalous in nature, such as chronic congestive cardiac failure or right atrial hypertension, for any reason. If such causes are eliminated, then enlargement of the coronary sinus results from an increased volume of flow of blood into the sinus through anomalous communications.

Enlargement of the coronary sinus may be divided into two broad groups based on the absence or presence of a left-to-right shunt into the coronary sinus.

Without Left-to-Right Shunt into the Coronary Sinus

When the coronary sinus receives an increased volume of systemic venous blood, several anatomic entities are possible. The most common condition is persistent left superior vena cava confluent with the coronary sinus. Less common conditions are those in which the coronary sinus anomalously receives veins of infradiaphragmatic origin. Examples of the latter include (1) partial anomalous hepatic venous connection with the coronary sinus and (2) continuity of the inferior vena cava with the hemiazygos vein, while the left superior vena cava is persistent.

Persistent Left Superior Vena Cava (Fig. 1a)

This is the most common thoracic venous anomaly and is likewise the most frequent cause of enlargement of the coronary sinus. While the true incidence of this anomaly is unknown, it has been stated\(^1\)\(^,\)\(^2\) that it occurs in from 3 to 10% of patients with congenital cardiac disease.

The left superior vena cava originates at the confluence of the left internal jugular and subclavian veins and passes ventrally to the aortic arch and root of the left lung. It then pierces the pericardium to become continuous with the coronary sinus. Winter\(^3\) reviewed

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Table 1

Classification of Anomalies Involving the Coronary Sinus

I. Enlargement of the coronary sinus
   A. Without left-to-right shunt into the coronary sinus
      1. Persistent left superior vena cava
      2. Partial anomalous hepatic venous connection to coronary sinus
      3. Continuity of inferior vena cava with left superior vena cava through hemiazygos vein
   B. With left-to-right shunt into coronary sinus
      1. Low pressure shunts
         a. Communication of coronary sinus with left atrium
         b. Pulmonary venous connection to coronary sinus
      2. High pressure shunts
         a. Coronary artery-coronary sinus fistula

II. Absence of coronary sinus
   A. Atrial septal defect localized to the position normally occupied by the coronary sinus
   B. Atrial septal defect involving entire lowermost portion of septum
      1. Persistent common atrioventricular canal
      2. Asplenia with congenital cardiac disease

III. Atresia of the right atrial coronary sinus ostium
   A. Persistent left superior vena cava
   B. Gross communication of coronary sinus with left atrium
   C. Multiple communications between coronary sinus and related atria

IV. Hypoplasia of the coronary sinus

Judging from the configuration of the P waves in the electrocardiogram, it has been suggested that when a persistent left superior vena cava is present, accessory sino-atrial nodal tissue may exist. This problem was studied histologically by Dr. Thomas N. James of the Henry Ford Hospital in a case in our files that was reported by Karnegis and associates. In that case the electrocardiogram suggested ectopic sino-atrial nodal tissue. In the specimen the right superior vena cava was absent while a persistent left superior vena cava was present and joined the coronary sinus in the usual manner. James’ observations, as yet unpublished, were that the sino-atrial node was normal and present in the usual position in the right atrium; that is, in close relationship to the anticipated entrance of the absent right superior vena cava. No sino-atrial nodal tissue was observed either near the coronary sinus or near the left superior vena cava.

Partial Anomalous Hepatic Venous Connection to the Coronary Sinus (Fig. 1b)

Nabarro studied a case in which a single large vein arising from the left lobe of the liver joined with a left phrenic vein to form

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Enlargement of the coronary sinus without left-to-right shunt into coronary sinus. (a) Persistent left superior vena cava confluent with coronary sinus (C.S.). (b) Partial anomalous hepatic venous connection with coronary sinus (modified from Nabarro). (c) Continuity of inferior vena cava with persistent left superior vena cava through the hemiazygos vein in a subject with situs solitus and multiple spleens. V.A. and V.V. = venous atrium and ventricle, respectively; A.A. and A.V. = arterial atrium and ventricle, respectively. (d) Continuity of inferior vena cava with persistent right superior vena cava through the hemiazygos vein in a subject with situs inversus and multiple spleens (c and d modified from Ongley and associates).
an anomalous channel. This vessel pierced the diaphragm and the pericardium. It then passed dorsal to the heart to join the coronary sinus. More recently, LePere and associates\(^7\) described an anomalous venous channel having a similar position and course within the pericardium. He did not, however, identify the infradiaphragmatic source.

**Continuity of the Inferior Vena Cava with the Left Superior Vena Cava through the Hemiazygos Vein (Fig. 1c and d)**

This condition occurs when the hepatic and prerenal segments of the developing inferior vena cava fail to fuse into a continuous channel. Under such circumstances, the prerenal segment of the "interrupted" inferior vena cava joins either the azygos or hemiazygos vein. The azygos vein drains into the right superior vena cava. If the inferior vena cava joins the hemiazygos vein, the latter, in turn, joins a persistent left superior vena cava. The left superior vena cava then is continuous with the coronary sinus.

It is recognized that when the inferior vena cava joins the right superior vena cava the coronary sinus is not enlarged. On the contrary, when the inferior vena cava joins the left superior vena cava (through the hemiazygos vein) the coronary sinus is enlarged, since the left superior vena cava joins the coronary sinus.

Continuity of the inferior vena cava with either superior vena cava may occur as an isolated benign anomaly. This condition has been described as incidental necropsy findings in persons up to 91 years of age.\(^8\) On the other hand, this condition may be associated with a wide range of cardiac anomalies, including cor biloculare, persistent common atrioventricular canal, anomalous pulmonary venous connection, atrial septal defect, pulmonary stenosis or atresia, or combinations of these.\(^1\) Other commonly associated anomalies include abnormal position of the heart, partial inversion of the abdominal viscera and polysplenia. Continuity of the inferior vena cava with the left superior vena cava was observed by one of us (J.E.E.) in another series, that of Ongley and associates.\(^6\)

In the latter series one case of normally positioned heart and polysplenia (case 3 of the Ongley series) showed the inferior vena cava to be on the left side and it joined the hemiazygos vein. The latter then terminated in a persistent left superior vena cava which, in turn, connected with the enlarged coronary sinus (fig. 1c). In this case, an atrial septal defect and total anomalous pulmonary venous connection to the right atrium were also present.

In another instance (case 4 of the Ongley series) a subject with total situs inversus and polysplenia showed the inferior vena cava to join the right-sided hemiazygos vein. The latter, in turn, entered the right-sided persistent superior vena cava which terminated in the enlarged coronary sinus (fig. 1d). In this subject there was partial anomalous pulmonary venous connection of the left pulmonary veins to the venous atrium.

**With Left-to-Right Shunt into the Coronary Sinus**

Highly oxygenated blood may be shunted into the coronary sinus through three possible anomalous communications. These communications may be divided into two groups depending on the pressure in the structure which makes anomalous connection with the coronary sinus. Low pressure left-to-right shunts occur as a result of anomalous communication of the coronary sinus either with the left atrium or with the pulmonary venous system. High pressure left-to-right shunts occur with fistulous communication between the coronary sinus and the coronary arterial system.

**Low Pressure Shunts**

Communication of the coronary sinus with the left atrium may be (1) indirect through a vein which bridges the two structures or (2) direct by way of an opening between the lateral extremity of the coronary sinus and the left atrial cavity. One case of each of these types was present in our series.

In a case previously reported by Eliot and associates\(^5\) a vein emerged from the left atrium, passed over the lateral wall of the chamber and terminated in the coronary sinus.
Enlargement of the coronary sinus with low pressure left-to-right shunts into coronary sinus. (a) Communication of the left atrium with the coronary sinus through an anomalous venous channel. Also present are stenosis of the right atrial ostium of the coronary sinus, anomalous connection of the left upper pulmonary vein to the left innominate vein, and a ventricular septal defect (modified from Eliot and associates9). (b) Gross communication between the left atrium and coronary sinus. (c) Total anomalous pulmonary venous connection to the coronary sinus and atrial septal defect. (d) Total anomalous pulmonary venous drainage resulting from anomalous pulmonary venous connection to the coronary sinus and to the left innominate vein.
sinus. The right atrial ostium of the coronary sinus was narrow (fig. 2a).

A fibrous strand extended between this vein and the left upper pulmonary vein. The latter connected anomalously with the left innominate vein. Also present in this case was a ventricular septal defect.

In the report dealing with this case the anomalous channel, termed a "levocardioarterio-cardinal vein," was considered to represent persistence of a primitive vessel connecting the left upper pulmonary vein and the left anterior cardinal vein. It was thought that this vessel persisted and developed in response to the partially obstructed ostium of the coronary sinus and functioned, during early development, as a collateral channel for egress of blood from the coronary sinus, the blood being carried into the developing pulmonary venous system. Later, as the involved pulmonary vein became incorporated into the left atrium, the resulting gross picture was that of a venous channel running between the left atrium and the coronary sinus. The direction of flow in this channel in the adult stage is unknown. Under usual conditions of pressure the flow would be expected to be in a left-to-right direction. In face of stenosis of the right atrial ostium of the coronary sinus, venous blood may have been carried into the left atrium through the channel.

A single case was observed in which an unusually large direct communication existed between the lateral extremity of the coronary sinus and left atrium (fig. 2b). This anomaly was found as an incidental condition in an elderly woman who died of causes unrelated to this anomaly. The magnitude of the shunt is not known but must have been considerable, as judged by the enlargement of the right atrium and ventricle.

**Pulmonary venous connection to the coronary sinus** exists in either total or subtotal forms. The more common form, total anomalous pulmonary venous connection to the coronary sinus, was observed in eight cases. This condition is characterized by each of the pulmonary veins joining a pulmonary venous confluence which, in turn, connects with the coronary sinus. An atrial septal defect or a valvular competent patent foramen ovale is regularly associated with this condition (fig. 2c).

We observed a single case of total anomalous pulmonary venous connection of unusual nature. The pulmonary venous confluence made two connections, one to the coronary sinus and the other to the left innominate vein (fig. 2d).

**High Pressure Shunts**

A *coronary artery-coronary sinus fistula* may involve either coronary artery to yield a high pressure left-to-right shunt into the coronary sinus. As is characteristic of arteriovenous fistulas, the involved artery is elongated, tortuous, and dilated (fig. 3a). According to the literature, such fistulas, when present, usually represent isolated anomalies. No cases of this specific type of coronary fistula were present in our series.

We observed a single case of coronary artery-coronary sinus fistula in a heart with aortic valvular atresia, hypoplasia of the left ventricle, and intact atrial septum (fig. 3b), a case reported by Raghib and associates. The only route for egress of blood from the left ventricle was through widely dilated myocardial sinusoids which connected with the left circumflex coronary artery. From the latter artery blood flowed in retrograde fashion into the coronary sinus through a fistula between the artery and the coronary sinus.

**Absence of the Coronary Sinus**

Absence of the coronary sinus is not known to occur as an isolated anomaly. It is regularly associated with other anomalies, namely persistent left superior vena cava terminating in the left atrium and atrial septal defect. Moreover, all hearts in which this anomaly is present have in common a right-to-left shunt at the left atrial level as a part of the functional abnormality.

When the coronary sinus is absent and a persistent left superior vena cava terminates in the left atrium, there are three anatomic subdivisions depending upon the type of
atrial septal defect and the presence or absence of additional anomalies. These will be considered in the immediately following material.

Atrial Septal Defect Localized to the Position Normally Occupied by the Coronary Sinus (Fig. 4a)

Raghib and associates,13 working in this laboratory, described three necropsy specimens in each of which there was a combination of the following anomalies: (1) termination of the persistent left superior vena cava in the left atrium, (2) absence of the coronary sinus, and (3) an atrial septal defect situated in the postero-inferior angle of the atrial septum, the position normally occupied by the coronary sinus. No other specific anomalies were associated, although in one of these cases a ventricular septal defect was present. The interesting functional effect of this complex is a left-to-right transatrial shunt associated with systemic arterial oxygen desaturation.

Defect Involving the Entire Lowermost Portion of the Atrial Septum

Persistent Common Atrioventricular Canal (Fig. 4b)

In contrast to the atrial septal defect described in the entity considered in the preceding section, the characteristic atrial septal defect in this group is more extensive. The defect occupies not only the postero-inferior angle of the atrial septum but also that part of the septum which characteristically is defective in persistent common atrioventricular canal.

Another characteristic of this group is that the atrioventricular valves show the characteristic malformations of persistent common atrioventricular canal (fig. 4b). The great arterial vessels are normally related, and the spleen is present.

The four cases of this subdivision of absence of the coronary sinus in our series are among 43 cases of persistent common atrioventricular canal being studied by Jue and associates.14
Congenital Cardiac Disease with Asplenia (Fig. 4c)

The final subdivision of absence of coronary sinus is that in which the anomaly is part of the developmental complex of splenic agenesis. This usually includes the following: (1) defects of, or absence of, the atrial and ventricular septa, (2) persistent common atroventricular canal, (3) pulmonary stenosis or atresia, (4) transposed great vessels, (5) anomalous connections of the pulmonary veins, and (6) bilateral superior vena cavae with absent coronary sinus.

In our series there are 15 examples of asplenia with persistent left superior vena cava joining the left atrium or the left side of a common atrium and absence of the coronary sinus. These were among the 17 cases of asplenia reported by Ruttenberg and associates. Usually, two large atrial septal defects are present which are separated by a strand of tissue, representing the only remnant of atrial septum.

Atresia of the Right Atrial Ostium of the Coronary Sinus

In this condition, the coronary sinus is present, but its opening into the right atrium is atretic. The coronary sinus lies in a normal position but ends as a blind sac. When a left superior vena cava is present, it functions as a collateral channel carrying blood in retrograde direction from the coronary system into the left innominate vein. From the latter, the blood is delivered into the right superior vena cava through which it flows into the right atrium.

Atresia of the right atrial ostium of the coronary sinus may occur as an isolated anomaly, although it has been reported in association with a number of other cardiac malformations. This anomaly is of little functional significance, although a right-to-left shunt of the coronary sinus blood into the left atrium occurs in some cases, depending upon the...
Atresia of the right atrial ostium of the coronary sinus. (a) The coronary sinus terminates as a blind sac and the left superior vena cava is persistent. Egress of blood from the coronary sinus is by retrograde flow through the left superior vena cava into the left innominate vein. Blood is then carried into the right superior vena cava and finally delivered into the right atrium. (b) Atresia of the right atrial coronary sinus ostium in which a narrow persistent left superior vena cava joins the coronary sinus. There is also anomalous communication between coronary sinus and left atrium. Major coronary sinus flow is probably into the left atrium rather than through the persistent left superior vena cava. (c) Atresia of the right atrial ostium of the coronary sinus. Blood from the coronary sinus drains into the related atria by way of enlarged thebesian veins.

Grant\textsuperscript{17} and Harris and associates\textsuperscript{18} described cases of atresia of the right atrial ostium of the coronary sinus in which the coronary sinus communicated with a persistent left superior vena cava. In the absence of a right atrial ostium, the route of egress of blood from the coronary sinus is in a retrograde direction, passing upward in the left superior vena cava, then into the right superior vena cava, by way of the left innominate vein, and eventually into the right atrium.

With Functional Persistent Left Superior Vena Cava (Fig. 5a)

With Gross Communication of Coronary Sinus and Left Atrium (Fig. 5b)

One of us (J. E. E.) observed in another series of cases an example of atresia of the right atrial ostium of the coronary sinus in which a narrow left superior vena cava joined the coronary sinus. In addition, the lateral extremity of the coronary sinus communicated with the left atrium in a manner somewhat similar to the communication illustrated in figure 2b. It is probable that the major flow from the coronary sinus was through this opening into the left atrium.

With Multiple Communications between Coronary Sinus and Related Atria (Fig. 5c)

We have observed three hearts in which there was atresia of the right atrial coronary sinus ostium and absence of the left superior vena cava. In this situation blood from the coronary sinus is carried into the related atria by means of enlarged thebesian veins.

Hypoplasia of the Coronary Sinus (Fig. 6)

This condition is seen when some of the cardiac veins fail to join the coronary sinus and empty individually into the atrial chambers through dilated thebesian channels. No major functional significance may be attributed to this condition of which we have observed three cases.
Comment

It is evident from the foregoing that among anomalies involving the coronary sinus there is a wide range of functional significance. Seldom is the coronary sinus the site of an isolated anomaly. In some situations there is no basic disturbance in the circulation, as in instances of persistent left superior vena cava with enlargement of the coronary sinus. In other situations significant functional disturbances result from the anomalous condition. In still other situations the circulation in the basic anomaly is fundamentally normal, but an alteration of the anomaly, such as interruption of an inferior vena cava that leads to a hemiazygos vein, may lead to significant systemic venous obstruction.

Observation that the coronary sinus is enlarged should raise the suspicion of flow from some anomalous source. The increased flow may result in troublesome effects during cardiopulmonary bypass. It is important that the possible sources be known lest essential anomalous structures, for example, anomalous hepatic veins or inferior venae cavae, be obstructed or sacrificed.

A point of interest to the surgeon is that an atrial septal defect situated in the postero-inferior angle of the atrial septum associated with absence of the coronary sinus should suggest the presence of a left superior vena cava terminating in the left atrium. Simple closure of such a defect will not correct the right-to-left shunt into the left atrium. Furthermore, if division of the left superior vena cava is contemplated, it should be borne in mind that the two superior venae cavae frequently are not joined by a left innominate vein. Raghib and associates make the point that in the absence of pulmonary hypertension, a left-to-right transatrial shunt associated with systemic arterial desaturation should suggest the presence of a left superior vena cava terminating in the left atrium, although this functional state may also occur when an atrial septal defect is located near the superior vena cava.

In instances of an unusually large communication between the left atrium and coronary sinus, catheterization data may be indicative of an atrial septal defect. In this condition, confusion may arise during operation, since viewed from the right atrium, there is no defect but only an unusually large coronary sinus. Treatment in such a case would consist of closure of the right atrial ostium of the coronary sinus.

Confusion may arise in cases of continuity of the inferior vena cava with the hemiazygos vein when a catheter is passed by way of a saphenous vein. In this instance the catheter may reach the right atrium by way of the hemiazygos vein, the left superior vena cava and finally the coronary sinus. Anderson and associates illustrated a case in which the cardiac catheter passed from the right superior vena cava to the inferior vena cava by proceeding through the following structures in the se-

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sequence given: right superior vena cava, right atrium, the coronary sinus, the left superior vena cava, the hemiazygos vein, and finally, the inferior vena cava.

Summary
A classification is presented of anomalies involving the coronary sinus. These anomalies are classified into four anatomic groups on the basis of (1) enlargement of the coronary sinus, (2) absence of the coronary sinus, (3) atresia of the right atrial coronary sinus ostium, and (4) hypoplasia of the coronary sinus. Anomalies involving the coronary sinus often are associated with other venous anomalies, either of the systemic or the pulmonary circulation. In some there is no basic disturbance of the circulation. Those conditions involving the coronary sinus which are of major functional significance participate in shunts, either left-to-right or right-to-left in nature. Enlargement of the coronary sinus in the absence of a shunt usually indicates that a systemic venous channel joins the coronary sinus anomalously.

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