

Inferior sinus venosus defect: Echocardiographic diagnosis and surgical approach

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Objective: We sought to define the inferior sinus venosus defect anatomically and document successful surgical approaches.

Methods: We identified all patients previously given a diagnosis of an inferior sinus venosus defect at the Hospital for Sick Children, Toronto, Canada, between 1982 and 2005 by interrogating the cardiology and cardiac surgery databases. We included those having interatrial communications in which 1 or more of the right pulmonary veins drained to the inferior caval vein but retained connection with the left atrium, the rims of the oval fossa, and the walls of the coronary sinus, both being intact.

Results: We identified 11 children who had an interatrial communication meeting the criteria for and undergoing surgical repair of an inferior sinus venosus defect. Median age was 1.2 years; 6 (55%) subjects were male, and none were cyanotic. Transthoracic echocardiographic analysis was performed preoperatively in all children, revealing right ventricular dilation in all. Surgical repair was accomplished with a pericardial patch. A complex baffle was needed in 3 children to maintain unobstructed inferior caval and pulmonary venous return. The echocardiographic diagnosis was complete in only 5 patients, but all diagnoses were correct since the year 2000. In all children the observations at surgical intervention showed that the defect was a venoatrial communication involving drainage of the right pulmonary veins to the inferior caval vein while retaining connection to the left atrium.

Conclusions: Transthoracic echocardiographic analysis should remain the modality of choice for diagnosis of the inferior sinus venosus defect. We report excellent surgical results with a patch or baffle, correctly redirecting the anomalous venoatrial connections.

The inferior sinus venosus defect is a rare congenital cardiac malformation outside the confines of the normal atrial septum produced as a consequence of anomalous pulmonary venous connection to the inferior caval vein while retaining the pulmonary venous connection to the left atrium.¹⁻³ Therefore rather than representing an atrial septal defect, the lesion is better considered in terms of an anomalous venoatrial communication. Such venoatrial communications have been well delineated with respect to the superior caval vein.^{1,4} Those involving anomalous connection to the inferior caval vein,² in contrast, are less well recognized. Thus far, diagnosis has been made either at the time of surgical repair or at autopsy. At present, there are no studies of large numbers of patients with such defects based on anatomic

diagnosis during life. Therefore we have sought to reinvestigate all patients identified with these defects during surgical correction at our institution. We have assessed the accuracy of the initial echocardiographic diagnosis, and we report the approaches used for surgical repair.

MATERIALS AND METHODS

Study Population

We identified all patients presumed to have an inferior sinus venosus defect at the Hospital for Sick Children, Toronto, Ontario, Canada, between July 1, 1982, and December 31, 2005. Our identification of such patients started with interrogation of the cardiology database to identify patients given diagnoses of an inferior sinus venosus defect. Of that group, there were very few who were coded correctly. Subsequently, given that all such patients undergo surgical intervention, we interrogated the surgical database. The initial approach was to cross-reference any child less than 18 years of age with an atrial septal defect, a partially anomalous pulmonary venous connection from the right lung, or both. This generated a list of 344 patients. From this list, we were able to identify 41 children in whom the surgical report described an interatrial communication in the environs of the inferior caval vein with or without partially anomalous pulmonary venous drainage. We examined the surgical reports of each of these patients to determine those patients with anatomy consistent with our understanding of the inferior sinus venosus defect, namely a venoatrial communication permitting shunting between the atrial chambers on the basis of anomalous pulmonary venous connection to the inferior caval vein with retention of the pulmonary venous connection to the left atrium. Using the surgical reports, we identified 11 children with this combination of findings. We then reviewed the medical records of these 11 patients. Ethical approval for the review was obtained from the Institutional Research Ethics Board of the Hospital for Sick Children.

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Measurements

We collected demographic data, clinical symptoms at the time of presentation, electrocardiographic indices, and diagnostic imaging modalities used. Echocardiographic analysis had been performed in all children preoperatively by using the transthoracic approach in all, with none undergoing a preoperative transesophageal echocardiogram. Echocardiographic measurements, diagnoses, and general descriptors for each patient were recorded from the detailed final reports.

Echocardiographic Data

It is our institutional policy that all children undergo a complete transthoracic echocardiogram as their initial study so as to reduce the likelihood of incomplete or inaccurate diagnoses. All patients with interatrial communications undergo systematic recordings of the short- and long-axial subcostal and parasternal views, the apical views, and the suprasternal views by means of both cross-sectional and color Doppler interrogation, from which a description of the size and site of each defect is recorded. There was no attempt to rereview these studies for accuracy. Instead, we chose to rely on real-time diagnostic accuracy, as recorded in the final report, in the absence of an established surgical or anatomic diagnosis (see below). For all patients, it was the anatomic information gleaned from the surgical database, the surgical notes, or both that was used as the gold standard for diagnosis.

Statistical Analysis

Descriptive statistical analysis was used to describe the characteristics of the patients and the surgical results as medians with ranges and means with standard deviations.

RESULTS

Anatomic Description

The inferior sinus venosus defect produces shunting between the right and left atrial chambers in consequence of anomalous pulmonary venous connection to the inferior caval vein, whereby the anomalous pulmonary vein or veins retain their connection to the left atrium. This produces a venoatrial communication (Figure 1) that overrides the intact rims of the oval fossa (ie, fossa ovalis). The normal atrial septum is made up of the floor and rims of the oval fossa, with the superior and posterior rims being infoldings of the atrial walls between the attachments of the caval veins to the morphologically right atrium and the right pulmonary veins to the morphologically left atrium. With the inferior sinus venosus defect, the pulmonary veins have retained their connection to the left atrium while also having achieved continuity with the inferior caval vein. If the pulmonary veins did not maintain their connection to the left atrium, the inferior caval vein would connect only to the right atrium, resulting in standard partially anomalous pulmonary venous connection to the inferior caval vein or inferior right atrium. It is because the pulmonary veins have attained the anomalous connection to the right atrium or its systemic tributaries yet retained their continuity with the posteroinferior wall of the left atrium that there is an extraseptal venoatrial confluence that permits the interatrial shunting. Therefore the essence of the inferior sinus venosus defect is a venoatrial connection of the pulmonary and inferior caval veins that is in continuity with both the right and left atrial walls.

The confluence thus created is outside the boundaries of the normal atrial septum; in other words, it is an extraseptal interatrial communication. As shown in Figure 1, the venoatrial communication is outside the confines of the oval fossa, which can itself be intact, be probe patent, or have a deficient floor permitting additional interatrial shunting. In the heart shown in Figure 1, A, which is held in the museum of Pittsburgh Children's Hospital, there is a persistent left superior caval vein draining to the right atrium through an enlarged coronary sinus. In the heart shown in Figure 1, B, from our own museum, there is also a persistent left superior caval vein opening to the coronary sinus, along with anomalous connection of the right superior pulmonary vein directly to the right atrium in the absence of a right superior caval vein. In both hearts the anomalous inferior venoatrial communication overrides the intact posteroinferior rim of the oval fossa. The association with persistent patency of the left superior caval vein is likely to be a chance finding. It should not be anticipated always to exist in patients with inferior sinus venosus defects.

Clinical Information

At the time of the operation, all the children included in our study were found to have comparable anatomic findings to the arrangement shown in Figure 1, with the exception of persistent patency of the left superior caval vein, which was not found in any of our patients. Of these patients, 6 (55%) were boys. At the diagnosis, median age was 1.2 years, with a range from 0.25 to 3.5 years. The initial median height was 81.9 cm, with a range of 61 to 102 cm, and the median weight was 9.9 kg, with a range of 5.9 to 16.0 kg. Blood pressure and heart rate were normal in all children. None of the children demonstrated cyanosis or desaturation as identified with the saturations of oxygen by means of pulse oximetry, this being recorded as between 97% and 100% in the most recent 6 patients for whom this test was available. All children had normal jugular venous pulsations, and none presented with respiratory distress. Hepatosplenomegaly was identified in only 1 child. As expected, all children demonstrated a widely split second heart sound and a systolic ejection murmur. Additionally, a middiastolic rumble was audible in 5 patients, with a right ventricular heave also palpable in 5 patients.

Diagnostic Tests

A scalar electrocardiogram was obtained preoperatively in 10 patients, all of whom had normal sinus rhythm. All children were found to have normal conduction, depolarization, and repolarization. By using voltage criteria, right atrial enlargement was diagnosed in 7 children, left atrial enlargement in 2 children, and right ventricular hypertrophy in 2 children, with none showing evidence of left ventricular hypertrophy. There were no instances of arrhythmia or any evidence of ischemic changes.

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