Atrial Septal Defects in Children
An Angiocardiographic Study

By John Lind M.D., and Carl Wegelius, M.D.

Atrial septal defects in children have been examined with an angiocardiographic technic which
permits conclusions concerning the dynamics of the heart. This method combines direct synchronous
biplane photography at a speed of 10 to 12 pictures per second with simultaneous electro-
cardiograms.

Defects in the atrial septum are of relatively great clinical importance. They are the most common of all
congenital cardiac malformations, constituting as a single lesion 7 to 25 per cent and as a
combined lesion perhaps as high as 85 per cent of all congenital cardiac anomalies. Moreover,
they are not infrequently accompanied by disturbances in circulation which cause symptoms sufficiently severe to compel the patient or the patient’s family to seek medical help. Except for an occasional instance, they are not as yet among those congenital cardiac lesions that are amenable to surgical treatment. Since these defects are not operable, their recognition is of importance in deciding whether a given congenital lesion can or cannot be treated surgically. With further surgical progress, possibly in the directions already being taken by Santy, Murray, and others, the sure recognition of atrial septal defects may become the prelude to their repair.

Diagnosis

At the present time the defects are often difficult to diagnose clinically, even with the aid of routine roentgenography. According to Taussig, among young adults atrial septal defect is the congenital malformation of the heart that is most frequently seen in medical practice and least frequently diagnosed correctly.

Needless to say, the paucity of specific clinical features is clearly reflected in the difference between the autopsy incidence of the lesion and the considerably lower clinical incidence. The first step toward further development must therefore be increased accuracy in diagnostic methods.

Previous Use of Angiocardiography

With the advent of angiocardiography it was hoped that a method had become available by means of which the diagnosis of atrial septal defect could be established with certainty. Unfortunately, however, such has not been the case. Despite the unquestioned value of angiocardiographic data in the evaluation of most congenital cardiac malformations, it has been of relatively little help in the diagnosis of atrial septal defects. Steinberg, Grishman and Sussman have been able to demonstrate defects by angiocardiography in only a small percentage of cases. Cosby and Griffith say simply that angiocardiography is “of little if any value” and Brown, Bing and de Groot apparently hold the same opinion.

Pathophysiologic Considerations

By direct measurement during heart catheterization in cases of atrial septal defect, it has been demonstrated that pressure in the left atrium usually exceeds that in the right atrium. Because of this pressure gradient the flow through an unguarded atrial septal defect is ordinarily from left to right. Not infrequently, however, it has been found that the pressure in the two atria is approximately the same and no shunt can be demonstrated. Furthermore, cases of atrial septal defect have

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been reported in which the direction of the shunt was mainly from right to left. These instances seems to be relatively rare except when associated with pulmonary stenosis or in terminal stages where cardiac insufficiency has been established. Soulé, Joly, Carlotti and Sicot\textsuperscript{41} have reported cases of mixed shunts; in this connection, it has been suggested that at some period in the cardiac cycle the shunt is reversed because of brief changes in the pressure gradient. The right atrium has been shown to fill faster than the left, and, because of this, it has been suggested that the right to left shunt takes place during atrial diastole. Many factors, however, appear to be involved, especially the atrial pressure variations incident to the respiratory cycle. The shunt in atrial septal defect seems to be bidirectional usually, a small amount of blood passing from right to left at a certain period of the heart cycle.

**Prerequisites for Diagnosis of Atrial Septal Defect by Angiocardiography**

A definite step forward seems to us to be offered by an altered angiocardiographic technic. We feel that, if we could make exposures rapidly enough in projections which give clear and free and, insofar as it is possible, ortho-roentgenographic visualization of the atrial septum, we might demonstrate graphically a brief right-to-left shunt. Even though there is no right-to-left shunt under usual circumstances, such a shunt may be produced by the altered pressure relationships incidental to the injection of the contrast medium. This would be more apt to occur in infants or children where the injection is made closer to the heart and the amount of contrast medium would result in relatively greater pressure differentials than in adults. Such a shunt can only take place during atrial diastole, as the orifices of the superior and inferior vena cava are functionally closed during atrial systole (fig. 4). Surely such a moment, in the first or second heart beat following arrival of the dye in the right atrium, would be the optimal time for demonstrating the defect. Relatively highly concentrated contrast medium from the right atrium should then pass through that defect into a left atrium which at this instant will contain no dye at all, and which will be visible since there will be no overlying shadows of dye in pulmonary vessels to confuse or to mislead.

Incidentally, it is at least possible that, with such a reversed pressure gradient as we assume might exist here for a split second, we might actually visualize a patent foramen ovale which under conditions of normal pressure would be functionally closed. In evaluating the clinical importance of such phenomena the amount of shunted dye is significant. It is evident that quite small persistent foramina lack pathologic importance, as indicated by routine roentgenographic data and clinical findings.

**Angiocardiographic Technic Used in Present Investigation**

The technic has as its essential feature synchronous roentgen photography in two planes at a rate of 10 to 12 exposures per second in each plane and with precise registration of the time of the exposures on a simultaneously recorded electrocardiogram.\textsuperscript{12}

The patient must be placed in such a position that in one projection the atria are not superimposed and the atrial septum lies as nearly as possible parallel to the course of the roentgen rays. We have found the two oblique views to be distinctly preferable to an anteroposterior and a lateral, the more informative of the two obliques being usually the left anterior. In the more commonly used anteroposterior projection, the atria overlie one another and it may be impossible to determine whether a shadow in this area is the result of opacification of the right, the left or both atria. The use of two projections simultaneously is, therefore, important and offers great help in interpreting the films by aiding the examiner to localize exactly the dye within the heart, to determine whether the dye is in the right atrium only or in the right ventricle as well and to determine the time of its first appearance in the left atrium. The usefulness of two simultaneously recorded projections has also been proved to us in cases of anomalous, supernumerary, left superior vena cavae in which a reflux of dye, recorded in one projection alone,
casts a shadow resembling that of an atrial septal defect.*

Investigations Carried Out

Studies of the Circulation through the Human Fetal heart

The course of the blood stream through the fetal heart has been a subject of controversy. The principal question has concerned the degree of separation or of mixing of the two streams entering the fetal right atrium, i.e., the well-oxygenated blood from the placenta and the poorly oxygenated blood brought by the superior vena cava. Recent studies in sheep fetuses have strikingly demonstrated the crossing of streams from caval veins with most of the blood from the placenta traversing the foramen ovale directly into the left atrium. Barclay and co-workers stressed, however, the difficulty of transferring the findings established in animal experiments to the human fetal circulation and the desirability of further angiocardiographic studies on human fetuses.

We have been able to perform angiocardiography on human fetuses obtained by legal abortions. The fetus was removed by cesarian section and as the angiocardiographic table was placed beside the operating table, the examination could be carried out within one-half minute.

When the opaque material is injected through the umbilical vein it can be followed through the ductus venosus directly into the right atrium. When the stream of dye enters the heart it divides on the free edge of the interatrial septum into a larger left stream and a smaller right stream, the former passing through the foramen ovale (fig. 1). In a series of 12 human fetuses aged 12 to 22 weeks this division of the blood from the inferior vena cava by a crista dividens could be demonstrated in all cases.

Clinical Consequences

The defects that are found in the interatrial septum are of varying congenital origin. Pathologists distinguish among patent foramen ovale, persistent ostium primum and persistent ostium secundum. It has been said that on clinical and physiologic grounds such distinctions are unwarranted. If an atrial septal defect exists, the direction and amount of shunt must depend upon the pressure relationships in the atria. Concerning the atrial septal defects, it may, however, be of interest to mention that in a series of five infants, where in each case angiocardiography was carried out twice by injection of dye through an antecubital vein and through a malleolar vein, the communication in three cases could only be shown by angiocardiography when the injection was made from below; that is to say, when the dye was introduced via the superior vena cava, no atrial patency could be shown (fig. 2), a result which corresponds with the actual demonstrated behavior of the fetal circulation discussed under the preceding heading. This result indicates that in cases of suspected atrial septal defect in infants which are to be studied by angiocardiography, the injection of contrast medium should be made from below.

The Closure of Foramen Ovale at Birth

It has long been known that shortly after birth the foramen ovale closes, thus completing the arrangement by which all the blood upon return from the body capillaries must go to the lungs before again starting on the systemic circuit. There is still a difference of opinion, however, as to how promptly the foramen closes.

With the expansion of the lungs the flow of blood through the lungs increases and more blood consequently returns to the left atrium, thus raising the pressure in this chamber. The

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Within these limits, the amount is governed by the cardiac volume.
Fig. 1. Entrance of blood from inferior vena cava into both atria during fetal life. Fetus aged approximately 21 weeks. Right anterior oblique position (RAO, a1-a3). Left anterior oblique position (LAO, b1-b3).

a1-b1: Dye passing through the umbilical vein and through the ductus venosus. a2-b2: Hepatic veins and inferior vena cava visualized. In entering the heart the dye is divided by crista dividens into both atria. a3-b3: One tenth of a second later, still no dye in the ventricles. The division is apparent. More dye in left than right atrium.
change in relative pressure thus produced in the two atria tends to close the valve of the foramen ovale and acts to keep it closed. The work of Barclay, Franklin and Prichard indicated that the contraction of the left atrium in itself closes the foramen ovale, and that this closure occurs physiologically immediately after birth and before that of the ductus arteriosus.

As the injection of contrast medium in human fetuses cannot be performed rapidly enough through the umbilical vein, because of the closure of ductus venosus, the state of patency of the foramen ovale and the ductus arteriosus must be studied in another way. In six newborn infants with no cardiovascular disease in whom angiocardiology was carried out by injection into the anteceubital vein within 12 hours after delivery, a diagnosis of atrial septal patency was made in two cases. In six other newborn infants where injection was done from below through a malleolar vein, also within 12 hours after delivery, the diagnosis of an atrial septal patency could be made in every case (fig. 3). The same amount of dye was used with both methods.

However, in all the cases investigated, where a patency had been proved, only insignificant amounts of dye were shunted into the left atrium.

After the sixth day of life, however, we have never been able to demonstrate any atrial septal patency in newborn infants (eight cases).

**Fig. 2. Demonstration of an atrial septal defect when dye was injected via inferior vena cava which was not shown when dye was injected via superior vena cava.** Left anterior oblique position and right anterior oblique position.

Injection of dye into a malleolar vein. No dye has passed into the ventricle at the beginning of the first visualized atrial systole. The stream of dye in the inferior vena cava is seen to be divided, a great part of it being directed into the left atrium through a septal defect, possibly as a result of mechanisms similar to that which is normal in fetal life (compare fig. 1).

**Functional Foramen Ovale in Asphyxia Neonatorum.** From the functional type of closure and the stage of development of the heart in the neonatal period it follows that the direction of the flow of blood may become reversed from the extra-uterine type back to the fetal type. That this happens not very infrequently seems to follow from the fact that we have observed several cases of asphyxia neonatorum in which angiocardiology demonstrated an intra-uterine type of circulation with the crista dividens directing most of the blood from the
inferior vena cava into the left side of the heart (fig. 4).

Angiocardiographic Findings in Atrial Septal Patencies in Infants and Young Children

Indirect Anatomic Evidence of Patency. Provided that the defect is sufficiently large, the indirect indications of atrial septal defect can be seen by fluoroscopy and routine radiologic examination. They can be demonstrated, often more strikingly, by angiocardiography. The heart is nearly always enlarged to the left and often to the right as well. The dilated right atrium displaces the distended right ventricle which, in turn, may form the rounded left border of the heart. Left atrium and ventricle
Fig. 4. Disturbances in the postnatal closure of foramen ovale in a case of anoxia in a newborn. Left and right anterior oblique positions. Injection into a malleolar vein.

a,b₁: The inferior vena cava heavily contrast-filled. Starting atrial contraction. Dye is stopped at the caval orifice. Retrograde contrast filling of the liver veins. About the same amounts of dye in right and left atrium. a,b₂: End of the following atrial diastole. A broad stream of dye can be seen passing over into the left atrium.
are of normal size or are small. The dilated pulmonary artery in contrast to the aorta is a prominent radiologic feature. In the left anterior oblique position the enlarged right ventricle is usually seen to project to the anterior chest wall, often displacing the left ventricle backwards. In the right anterior oblique position the esophagus is seen to be diffusely pushed backward by the large heart, though the left atrium is not enlarged.

Indirect Hemodynamic Evidence of Patency. The profuse pulmonary vascularity is often seen in both oblique projections. Having the same significance as the hilar dance, often seen fluoroscopically, is an increased amplitude of the pulsations of the pulmonary artery.

A persistent opacification or reopacification of the right atrium and ventricle and the pulmonary arteries from the left heart gives presumptive evidence of interatrial communication, with a left-right shunt. This conclusion is justified only when it can be established that the dye in the right heart can have come only from the left heart. If the dye is injected slowly or if its disappearance from an afferent vein is delayed—a not infrequent event when the dye is carried to the heart via the inferior vena cava—the appearance of persistent opacification or reopacification is produced, the source of the dye obviously being the venous tributaries of the right heart and not the left heart. In the left anterior oblique position persistent opacification of the right side of the heart can be identified clearly. In the right anterior oblique position it can be established that the right atrium and not only the right ventricle and pulmonary artery are opacified. This is of some differential importance since, with interventricular septal defect, only the right ventricle and pulmonary artery show persistent opacification or reopacification.

As a consequence of the shunt the visualization of left ventricle and aorta is generally poor. The individual signs are well known and need not be described in detail or illustrated here.

Direct Demonstration of Atrial Septal Patency

In view of the considerations which have been discussed, it seemed to us that, by a modified technic, it might be possible to demonstrate a direct spread of contrast substance from the right to the left atrium during the first few heart cycles after the injection, irrespective of the direction of the interatrial shunt. We now wish to discuss the results of our studies.

Material and Results. A review of our material consisting of 150 certain or suspected cases of congenital heart disease studied by angiocardiography revealed that in 30 cases (20 per cent) the atrial septal patency has been distinctly visualized. In many cases it has further been possible to make observations concerning the size, localization and pathophysiology of the patency.

In comparison it may be mentioned that in a smaller series of 12 infants less than 2 years of age and without evidence of cardiovascular disease none showed evidence of atrial septal patency.

Size of the Septal Patency. Despite the embryologic and anatomic interest one may find in the many different forms which atrial septal defects may display, the clinical importance of those defects depends, according to Roesler, not upon their form or their position but rather upon their size.17 He suggested that only those defects which measure at least 1 cm. in diameter are clinically significant. Such a standard applied to infants and children would, however, obviously be unsatisfactory because of the variable growth of the defect along with the growth of the heart, and, particularly, because it seems most reasonable to assume that clinical importance depends not so much upon absolute size of the defect as upon its relative size, relative, that is, to heart size. In connection with this it may be pointed out that the size of the patency in the working heart probably is not constant. Several observations we have made seem to show that the size of a defect can change during the heart cycle and that the opening is smallest at the height of atrial systole (fig. 5).

It has been possible to recognize by angiocardiography the size of the patencies directly and thus to rule out—at least in most cases—the possibility of the presence of a simple patency of the foramen ovale, or small and
perhaps clinically inconsequential septal defects.

Localization. In most cases the left anterior oblique view gives some information about the localization of the defect. The possibility that the localization of the defect may occasion peculiarities in the clinical picture has already been mentioned.

Pathophysiology. If the pressure is higher in right atrium than in the left—for instance in cases associated with pulmonary stenosis—the right-to-left shunt of dye is most marked during atrial systole; no prolonged opacification or reopacification of the right atrium appears at the end of the examination.

If the dye is shunted into the left atrium mainly during atrial diastole, it may be due to the existence of bidirectional shunt or a reversal of the shunt caused by the increased pressure in the right atrium incident to the injection of the contrast medium. This can be confirmed by later pictures of the series showing the shunt of dye back from the left atrium into the right mainly during atrial systole. This shunt is diagnosed either by a maintained opacification of the right heart or by a real reopacification. In cases of open foramen ovale and small atrial septal defects no such shunt of the dye from left to right atrium will be seen.

If there is a big shunt from left to right, the dilution of dye in the right atrium and ventricle through blood shunted from the left atrium becomes apparent in the series of pictures as long as the concentration of dye is much higher in the right than in the left atrium.

It has been pointed out that the findings of persistent opacification (actually reopacification) of the right atrium, right ventricle and pulmonary arteries must be interpreted with extreme caution. Caution is particularly necessary if an angiocardiographic method used consists in making 1 to 2 exposures per second in one projection only. The interpretation is, however, easier and can be more accurate if there is available for study a series of films in which are included synchronously recorded views in two projections and in which the interval between successive exposures is sufficiently brief to permit a continuous registration of the passage of the dye through the heart.

In pathologic cases in which the function of the right atrium is interfered with, systolic reflux of dye into the vena cavae is very common (fig. 6). In cases of atrial septal defects with significant left-to-right shunt, reflux most often into the inferior vena cava, is regularly seen. As the closure mechanism which sur-
At this instant of the cardiac cycle, the orifice of the vena cava is closed and the dye is prevented from entering the atrial cavity, the result of this being that the venous tributaries of the vena cava become filled in retrograde fashion with the contrast medium.

**SUMMARY**

The clinical importance of atrial septal defects is pointed out. It is recalled that angiocardiography has previously been considered of relatively little help in the diagnosis of this malformation. Studies are reported which show...
that the fault has been with the technic of its application rather than with the method itself. The positive diagnostic value of a right-to-left shunt which can be demonstrated by a special angiocardiographic technic is stressed.

**Sumario Español**

Defectos del septo atrial en niños han sido examinados por medio de una técnica angiocardiográfica que permite conclusiones concernientes a la dinámica del corazón. Este método combina fotografía sincrona bipolar a una velocidad de 10 a 12 placas por segundo con electrocardiograma simultáneo.

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