Auscultatory Recognition of Aneurysm of the Membranous Ventricular Septum Associated with Small Ventricular Septal Defect

By Daniel R. Pieroni, M.D., Barbara B. Bell, M.D.,
L. Jerome Krovetz, M.D., Ph.D., P. Jacob Varghese, M.D., M.R.C.P.,
and Richard D. Rowe, M.D., F.R.C.P. (Edin.)

SUMMARY
The clinical diagnosis of aneurysmal formation of the membranous ventricular septum associated with a small ventricular defect has been made previously only by means of angiocardiography. Active movement of the aneurysm during cineangiocardiography suggested the possibility of a corresponding auscultatory event. When careful auscultatory and phonocardiographic examinations were performed on 21 patients previously documented as having a small membranous ventricular defect with an associated aneurysm, a distinct early systolic sound was heard in 17 (81%). This sound was "clicky" in quality, confined to a narrow area along the lower left sternal edge, and best heard in expiration. It occurred during the upstroke of the carotid arterial tracing and followed the Q wave by 100 to 130 msec.

On the basis of experience with other patients the development of this early systolic sound in a patient with a small ventricular septal defect suggests the diagnosis of associated septal aneurysm, but such a sequence requires confirmation by serial angiocardiography. Aneurysmal formation may be a prelude to spontaneous closure of the septal defect in which case auscultation of an early systolic sound should not only prove to be a valuable diagnostic sign for the clinician but may also be of considerable prognostic significance for the patient.

Additional Indexing Words:
Aneurysm of ventricular septum
Pediatric study
Spontaneous closure of ventricular septal defect
Auscultation
Early systolic sound
Phonocardiography

In 1957 Steinberg demonstrated an aneurysm of the membranous ventricular septum by angiocardiography. So far this has been the only means of making the clinical diagnosis. Since cardiac catheterization and angiocardiography still involve some risk to the patient, a noninvasive technique is desirable to confirm the presence of an aneurysm of the membranous septum in association with a small ventricular defect.

Distention and relaxation of the aneurysm during the cardiac cycle when viewed on the cineangiocardiogram led us to suspect that a

From the Department of Pediatrics, Johns Hopkins University and The Helen B. Taussig Children's Cardiac Center, The Johns Hopkins Hospital, Baltimore, Maryland.

Supported by U.S. Public Health Service grant 5R01 HE10718 and The Maternal and Child Health Service Project grant 12 H201.

Dr. Krovetz is the recipient of a Research Career Development Award (5K03-HE-09761-05).

Presented in part at the 43rd Annual Scientific Sessions of the American Heart Association, Atlantic City, New Jersey, November 14, 1970.

Address for reprints: Daniel R. Pieroni, M.D., The Helen B. Taussig Children's Cardiac Center, Johns Hopkins Hospital, Baltimore, Maryland 21205.

Received January 18, 1971; revision accepted for publication June 18, 1971.

Circulation, Volume XLIV, October 1971 733
corresponding auscultatory event might be present. When subsequent careful auscultation of these patients was performed a distinct early systolic sound was commonly noted. This paper describes the detailed auscultatory and phonocardiographic observations in 21 children with small ventricular septal defects and aneurysms of the pars membranacea.

Methods

Thirty-six patients with isolated membranous ventricular septal defects underwent cardiac catheterization in the Pediatric Cardiovascular Diagnostic Laboratories of the Johns Hopkins Hospital between August 1967 and September 1969. The techniques employed to document the diagnosis included blood oxygen determinations and indicator-dilution curves followed by left ventricular angiography in the left anterior oblique position. All were assigned as having small (mild) ventricular septal defects according to the criteria outlined by the National Natural History Study (Lambert, EC: Personal communication): namely, Qp/Qs < 1.4 and Pp/Ps < 0.5. Fifteen of the 36 patients (42%) had an isolated small ventricular septal defect of the ordinary type and 21 (58%) had an aneurysm associated with the septal defect. The group showed an equal ratio of males to females. The age range was 3.1 to 12.8 years. All patients were recalled for phonocardiographic examination from 0.4 to 3.4 years following catheterization.

Detailed auscultation was performed on each child by at least two of us with specific focus upon early systolic events.

Phonocardiograms were taken with the patient in the recumbent position using a polybeam recording system* (350-1700C heart sound preamplifier, 350-3200 A ECG preamplifier and 4560 recorder). Tracings were taken at a paper speed of 100 mm/sec with 0.04-sec time lines using low end cut-off frequencies of 50, 100, and 200 Hz through two contact crystal microphones placed simultaneously at the second left intercostal space (LICS) and the apex, second right intercostal space (RICS) and fourth LICS, second and fourth LICS, third and fourth LICS, and over the precise area where the early systolic sound was detected. In addition to an electrocardiogram and apexcardiogram, the respiratory phases, indirect carotid arterial and jugular venous pulses were recorded as points of reference. The interval between the Q wave of the electrocardiogram and the early systolic sound (Q-ESS) and the interval between the Q wave and the first heart sound (Q-1) were measured to delineate the temporal relationships of intracardiac events.7

Results

An early systolic sound was noted on auscultation in 19 of the 21 patients with an associated ventricular septal aneurysm and in only one of the 15 without an aneurysm. The early systolic sound which occurred shortly after the first heart sound, was “clicky” in quality and usually was not loud. Although it was best heard in expiration, the timing in the cardiac cycle remained constant in all phases of respiration. This sound was usually localized to a small area along the lower left sternal border or occasionally near the cardiac apex. It rarely had wide radiation.

All patients had a holosystolic murmur found in the usual left sternal area (fig. 1). In addition several patients had late systolic accentuation of the murmur in rather localized but not always consistent areas.

The phonocardiograms of 17 (81%) of the 21 patients with an aneurysm confirmed the presence of an early systolic sound (fig. 2) with a Q-ESS range of 100 to 130 msec (mean, 118 msec). An early systolic sound was recorded in only one of the 15 patients without an associated aneurysm. In all cases the early systolic sound occurred on the upstroke of the carotid tracing and at or shortly following the E point of the apexcardiogram (fig. 3). Gauged by the intensity of the first sound, the early systolic sound was loud in three patients, of medium intensity in five, and soft in nine. It was recorded on mixed frequencies. The Q-1 interval range was 40 to 70 msec (mean, 53 msec).

Cardiac catheterization and left ventricular angiocardiography showed that all 36 patients had a defect in the membranous portion of the ventricular septum, 21 having an associated aneurysm of the pars membranacea. Only six patients of the entire group had a peak difference in systolic pressure across the pulmonary valve greater than 5 mm Hg (maximum, 10; mean, 6 mm Hg). Three patients had a peak systolic pressure difference across the aortic valve greater than 5 mm

*Hewlett-Packard, Waltham, Massachusetts.
AUSCULTATORY RECOGNITION OF ANEURYSM

Phonocardiograms (PCG), indirect carotid artery pulse, and electrocardiogram (ECG). Typical tracings taken in a patient without an aneurysm. Note the holosystolic murmur with normal first ($S_1$) and second ($S_2$) heart sounds in the absence of an early systolic sound (ESS).

Figure 1

Hg (maximum, 8; mean, 6 mm Hg). No patient had evidence of atrioventricular valve insufficiency. Angiographically the aneurysms varied in length, neck width, and number of distal apertures. The most common type of aneurysm had a short, wide neck with either single or multiple distal apertures (fig. 4).

Discussion

In a recent comprehensive review of the literature, Hamby and colleagues assembled 136 aneurysms of the membranous septum. Ninety-eight were confirmed by autopsy, 27 by angiography, and 11 at surgery. The relatively high incidence of aneurysmal formation in our series probably resulted from the fact that small asymptomatic ventricular septal defects are not usually investigated by left ventricular angiography.

These peculiar defects are almost always associated with small-sized interventricular communications and have not been diagnosed by external examination in previous reports. An early systolic sound was substantiated in 17 (81%) of the 21 patients with an aneurysm of the membranous septum while it was recorded in only one of 15 patients with the isolated septal defect without an aneurysm. Therefore, when an early systolic sound is present in a patient with a small membranous ventricular septal defect, there is a high probability of an associated aneurysm.

On auscultation the early systolic sound was localized predominantly along the lower left sternal border and occasionally at the apex. It did not radiate, was always heard in early systole, and was most easily detected during expiration. The location of the early systolic sound and its radiation were essential qualities.
Phonocardiograms (PCG), indirect carotid artery pulse, and electrocardiogram of a patient with an aneurysm and an audible early systolic sound (ESS). The ESS is of high intensity. A holosystolic murmur (SM) occurs along the lower left sternal border with normal first (S₁) and second (S₂) heart sounds.

in separating this sound from high intensity, high frequency, more widely radiating aortic ejection clicks. These same features were also of value in differentiating the early systolic sound from pulmonic ejection clicks which are localized to an area higher along the left sternal border. The Q-ESS, Q-aortic and Q-pulmonic ejection click intervals frequently overlap and, therefore, are not of value in differential diagnosis. The temporal relationship of the early systolic sound to the electrocardiogram, apexcardiogram, and carotid and jugular pulses distinguished this sound from split first heart sounds, late systolic clicks, and fourth heart sounds.

It was necessary to maintain a high index of suspicion when examining patients with a small ventricular septal defect since the early systolic sound might easily be overlooked on superficial auscultation. Various problems arose in documenting the existence of the early systolic sound. Besides time and patience, phonocardiographic confirmation of the sound required careful positioning of the microphone following detailed auscultation, correct band phasing, and recording in expiration. Documentation of the sound was not possible when ideal conditions were not attained. This was thought to be the reason why we were unable to record the sound in two patients who were noted to have an early systolic sound on auscultation.

The murmur in all patients was holosystolic. In several patients with an associated aneurysm, late systolic accentuation of the murmur was noted. This particular finding has been reported previously, and, if it is not actually unique to, may be common in small
Figure 3

Two tracings taken in the same patient. (Upper) The ESS occurs on the upstroke of the indirect carotid artery pulse. (Lower) The ESS corresponds to the E point of the apexcardiogram (ACG).
ventricular septal defects with an associated aneurysm.

Several possible explanations may account for the origin of the early systolic sound. It may arise either from collisions or jet streams between the aneurysm and the tricuspid valve or ventricular wall. This concept has been invoked by some investigators as a cause of ejection clicks. On the other hand, since either a normal or pathologic tricuspid valve may partially occlude a membranous defect so that it resembles an aneurysm, it is conceivable that an accentuated delayed tricuspid closure sound may result. A similar mechanism of delayed closure has recently been recorded in patients with Ebstein's anomaly.

Although we have been unable to relate the sound to size, shape, or specific angiographic appearance of the aneurysm, we are inclined to favor the possibility that the early systolic sound is produced by the valvelike tensing of the aneurysm at the time of peak filling, a mechanism previously shown to be responsible for aortic ejection clicks.

Pombo and coworkers reported finding an ejection click in a patient with an aneurysm of the membranous septum and a systolic pressure difference of 10 mm Hg across the pulmonary valve. They assumed that the click was secondary to valvular stenosis. Since we have recorded an early systolic sound in 12 patients without significant systolic pressure difference across the pulmonary valve, it seems likely that whatever the underlying mechanism may be, it is not due to valvular stenosis or dilatation of the pulmonary artery.

One patient in our series is of special interest. Before catheterization, a systematic search for an early systolic sound by auscultation and phonocardiogram was negative. As predicted the angiocardiogram revealed a small membranous ventricular septal defect without an aneurysm. One and one-half years later an early systolic sound was noted for the first time on auscultation and recorded on the phonocardiogram. On the basis of our experience with other patients the development of an early systolic sound in this patient suggests that he has developed an aneurysm although angiocardiographic confirmation has not yet been obtained.

Several reports suggest that aneurysmal formation may be the prelude to or a mechanism for spontaneous closure of small membranous defects. If this sequence of events can be confirmed, then the detection of an early systolic sound in a patient with clinical evidence of a small ventricular septal defect may prove to be a valuable diagnostic aid for the clinician and an important prognostic indicator of future spontaneous closure.

Acknowledgment

The authors wish to extend their appreciation for the skillful technical assistance of Mrs. L. Rogers and the help given during preparation of the manuscript by Dr. L. Haroutunian and Mrs. D. Mitchell.

References

1. Steinberg I: Diagnosis of congenital aneurysm of the ventricular septum during life. Brit Heart J 19: 8, 1957

Circulation, Volume XLIV, October 1971
AUSCULTATORY RECOGNITION OF ANEURYSM

Auscultatory Recognition of Aneurysm of the Membranous Ventricular Septum Associated with Small Ventricular Septal Defect
DANIEL R. PIERONI, BARBARA B. BELL, L. JEROME KROVETZ, P. JACOB VARGHESE and RICHARD D. ROWE

_Circulation_. 1971;44:733-739
doi: 10.1161/01.CIR.44.4.733
_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1971 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/44/4/733

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in _Circulation_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation_ is online at:
http://circ.ahajournals.org//subscriptions/