Aneurysm of the membranous septum in adult patients with perimembranous ventricular septal defect

Ahmet T. Yilmaz*, Ertugrul Özal, Mehmet Arslan, Harun Tatar, Ömer Y. Öztürk

Department of Cardiovascular Surgery, Gülhane Military Medical Academy, Ankara, Turkey

Received 25 January 1996; revised 15 May 1996; accepted 23 May 1996

Abstract

Objective: The aneurysm of the membranous septum (AMS) has often been considered as benign in the minds of many previous investigators. We have analyzed the complications associated with AMS in adult patients. Methods: Fifty-one cases (20%) of AMS in 254 adult patients with perimembranous ventricular septal defect (VSD) are described. The diagnosis of AMS was based on angiographic criteria. Thirty-nine (76.5%) of the 51 patients with AMS were aged between 20 and 29 years. All patients but one with AMS had a pulmonary-to-systemic flow (\(Q_p/Q_s\)) of less than 2.3 (range 1–2.1, mean 1.4). In a patient who had a ruptured aneurysm, the \(Q_p/Q_s\) was 2.7. There were six main complications affected by AMS and/or VSD: aortic valve prolapse in 24 patients (47%), aortic regurgitation in 15 (29.4%), tricuspid insufficiency in nine (17.6%), right ventricular outflow tract obstruction in two (4%), and rupture of the aneurysm in one patient (2%). Seven patients (13.7%) had prior bacterial endocarditis. All patients underwent surgery. Aneurysm and VSD were closed by direct suture in nine and with a patch in 42 patients. Aortic valve repair was performed in 13 patients in whom regurgitation was mild to moderate, and replacement was required in two patients with severe aortic regurgitation. Results: There were no early or late deaths. Residual communication and recurrence of the aneurysm was noted three and seven years postoperatively in two patients where VSD had been closed by direct suture. Conclusions: According to present data, aneurysm formation functionally reduces the VSD size, but it has the potential consequence of promoting tricuspid insufficiency, aortic valve prolapse, right ventricular outflow tract obstruction, rupture and bacterial endocarditis. Therefore, we recommend that AMS should be resected completely and the defect produced closed with a patch in order to prevent further enlargement and consequent complications even if there are no cardiac symptoms. © 1997 Elsevier Science B.V.

Keywords: Aneurysm of the membranous septum; Ventricular septal defect; Aortic regurgitation; Bacterial endocarditis; Tricuspid insufficiency

1. Introduction

The aneurysm of the membranous portion of the ventricular septum (AMS) has been an interesting and controversial subject for many years. A hemodynamically large membranous ventricular septal defect (VSD) in infancy progresses to a functionally smaller defect with aneurysm formation later in childhood. Our present knowledge of the clinical and hemodynamic consequences of AMS has mainly derived from studies of children. However, the actual incidence and natural history of AMS associated with VSD has not been well documented in adult patients. We have analyzed the incidence, clinical and hemodynamic findings and complications in our patients with AMS who had reached adulthood at the time of diagnosis.
2. Materials and methods

The study population comprised the total number of patients seen in the years 1980–1995 who were over the age of 15 years when the diagnosis of isolated perimembranous VSD was made. The diagnosis of VSD was based on findings during right–left catheterization and angiogram. AMS was defined as angiographic evidence of any alteration or deposition of tissue around the margins of a membranous VSD, producing a structure with distinct margins which protrudes into the right ventricle with each systole and seems to partially or completely occlude the flow of the blood through the associated defect [6] (Fig. 1). In 51 (20%) of 254 patients with perimembranous VSD, AMS was demonstrated by angiographic criteria described. Anatomic evidence of AMS was examined during the operations. Membranous septum tissue was defined as a well-developed, thickened fibrous tissue, perforated by one or many holes, forming an aneurysm of the membranous septum that bulges forward into the right ventricle. It was unattached to the tricuspid valve and not part of the tricuspid valve mechanism. When the AMS associated perimembranous VSD resulted in adherence of the leaflet tissue to the edges of the defect, this pathology was clearly defined by the tricuspid leaflet itself and chordal attachment.

The 254 patients comprised 31 women and 223 men. Fig. 2 gives the age distribution of patients with VSD and AMS. Thirty-two (62.7%) of the 51 patients with AMS were symptomatic and 19 were asymptomatic. The severity of the VSD according to hemodynamic criterias was assessed using the criteria of the natural history study [12] (Table 1). The degree of aortic regurgitation was determined by aortic root angiogram, and graded as mild, moderate or severe according to standard criteria [2].

All patients with AMS underwent surgery. Attempts for AMS isolation were performed by the transatrial approach or right ventriculotomy. Routinely, we have emphasized opening the tricuspid valve in a radial fashion about 2 cm wide to expose the true perimeter of the defect and aneurysm during the transatrial approach. After the area of the VSD was exposed, the aneurysmal sac was identified and a radial incision was made in the aneurysmal sac. Great care was taken not to injure the tricuspid valve. Then, the aneurysm was excised at its edges unless it was atypical and part of the tricuspid valve mechanism. The edges of the defect proper were then clearly visible. Double-armed sutures buttressed with pledgets were placed around the perimeter of the VSD, passed through the edge of an appropriately sized Dacron patch, and then tied, securing the patch against the defect. When the septal leaflet of the tricuspid valve covered the VSD as a saclike form structure that adhered to the margins of the defect, no attempt was made to excise the aneurysmal sac. In these cases, VSD was repaired in the usual way, and then, the edges of the incision in the tricuspid valve leaflet were approximated with interrupted sutures. In the aortic valvuloplasty, the redundant portions of the cusps were plicated and fastened against the aortic wall with pledgetted matress stitches.

![Fig. 1.](image1.png) Fig. 1. Left ventricular angiography in the left anterior oblique projection showing the aneurysm of the membranous septum in a 29-year old man. A ventricular septal defect forms the base of the aneurysm, while contrast enters the right ventricle through a trivial defect at its apex ($Q_p/Q_s = 1.2$). The aneurysm protruded into the right ventricle outflow tract, and showed a pressure gradient of 32 mmHg across the outflow tract of right ventricle.

![Fig. 2.](image2.png) Fig. 2. Age distribution in 254 adults with a late diagnosis of congenital isolated perimembranous VSD and AMS.

### Table 1

<table>
<thead>
<tr>
<th>Definition of severity for ventricular septal defect [12]</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD</td>
</tr>
<tr>
<td>------------------</td>
</tr>
<tr>
<td>Mild</td>
</tr>
<tr>
<td>Moderate</td>
</tr>
<tr>
<td>Severe</td>
</tr>
<tr>
<td>Inoperable</td>
</tr>
</tbody>
</table>

PA, pulmonary artery; AO, aorta; $Q_p$, pulmonary flow; $Q_s$, systemic flow; $R_p$, pulmonary resistance; $R_s$, systemic resistance.
3. Results

Angiographic evidence of aneurysm was found in 20% of 254 perimembranous VSDs. Thirty-nine of the 51 patients with AMS (76.5%) were between 20 and 29 years of age (Fig. 2). Fig. 3 presents the distribution of severity of the perimembranous VSD and notes the incidence of AMS in each severity category. All patients with AMS but one had a ratio of pulmonary-to-systemic flow ($Q_p/Q_s$) of less than 2.3 (range 1.1–2.1, mean 1.4). Nineteen patients (37.2%) with AMS had a $Q_p/Q_s$ of 1.5 or greater. In 32 patients, the $Q_p/Q_s$ was under 1.5. Eisenmenger’s disease was not found in any of the 51 patients with AMS, but was present in two patients (0.99%) of the 203 perimembranous VSDs without AMS.

Six patients with AMS had Down syndrome and five of these patients had a $Q_p/Q_s$ of 1.5 or greater. There were coexistent cardiac defects including valvular pulmonary stenosis in three patients, coarctation of aorta in two patients, secundum type atrial septal defect in five patients, partially anomalous pulmonary venous connection in one patient, and persistence left superior vena cava in two patients.

There were six main complications affected by AMS and/or VSD; aortic regurgitation, bacterial endocarditis, right ventricular outflow tract obstruction (RVOTO), tricuspid insufficiency, and rupture of the aneurysm (Table 2). Rupture of the aneurysm was found in one patient of whom an angiography had previously shown a large aneurysm with an insignificant $Q_p/Q_s$ and normal pressures. After three years, the patient returned with sudden onset symptoms of congestive failure. Recatheterization revealed severe shunt with a $Q_p/Q_s$ of 2.7, and was accompanied by aortic valve prolapse and mild aortic valve regurgitation. Two patients had a chronic supraventricular tachycardia. There was a right bundle branch block in five patients, and left bundle branch block in one patient. Nine patients (17.6%) with AMS had angiographic evidence of tricuspid insufficiency with no clinical symptom of such. In each case, contrast material was observed to enter the right atrium following a selective left ventricular injection. None of our cases showed evidence of a defect in the atrial portion of the membranous septum. In these patients, aneurysm formation was involved in the apposition of tricuspid septal leaflet tissue. Two patients had a significant RVOTO (32 mmHg and 24 mmHg gradient) which could be attributed to the presence of an aneurysm, as RVOTO disappeared after operation for only AMS in these patients. In these patients, there were large, long aneurysms of the wind sock variety.

Seven (13.7%) patients had a history of bacterial endocarditis, and one had experienced two episodes. Two had bacterial endocarditis several years before admission, and in five this complication had occurred within the past year. The calculated values of $Q_p/Q_s$ in these patients were under 1.5 and patients had normal pulmonary pressure. There was a left ventricle to right atrial (LV-RA) shunt in three of seven patients with bacterial endocarditis.

Twenty-four (47%) patients had aortic valve prolapse (AVP), and aortic regurgitation was found in 15 (29.4%) patients. Eight patients had prolapse of the noncoronary aortic cusp, and others had prolapse of the right aortic cusp. There was mild aortic regurgitation in 11, and moderate to severe aortic regurgitation in four of them. In two patients with severe aortic regurgitation, aortic valve replacement was required, as not only was there a prolapsing of aortic cusps, but also secondary aortic annular dilatation and cusp retraction.

All patients with AVP except three had small $Q_p/Q_s$ values (<1.5). In three patients, the $Q_p/Q_s$ was 1.5 or greater. The incidence of aortic regurgitation and aortic valve prolapse was significantly higher in patients with AMS (29 and 47%) than patients with VSD (4.9 and 20.2%). There was a mild shunt ($Q_p/Q_s < 1.4$) in 58 of 65 patients with aortic valve prolapse associated with VSD or AMS.

During the initial period of this study, the right ventricular approach together with transatrial approach was used for the repair of VSDs in 11 cases. Recently, repair was performed through a right atrial approach only in nearly all cases (40 cases). Defects were closed.
by direct suture in nine patients. Residual communi-
cation and recurrence of the aneurysm was noted
postoperatively in two of these patients. In other pa-
tients, routine aneurysms (adjacent tissue onto the
VSD except tricuspid valve mechanism) were excised
completely and the created large defects were closed
with a Dacron patch. In all closures with a patch,
there were no reoperations for residual or recurrent
VSD and/or AMS. During the surgical examination,
the origin of the aneurysm was confirmed; in six
cases, the aneurysm was entirely formed by adher-
ence of the tricuspid septal leaflet tissue; in eleven
cases, the aneurysm was composed of the tricuspid
valve leaflets and interventricular component of the
membranous septum tissue; and in another 28 cases,
possessed membranous septum tissue only. In six pa-
tients the origin of the aneurysm was not confirmed.
Aneurysms were usually small to moderate size,
varying from 1–3 cm in diameter. In two cases,
there were large aneurysms, and aneurysmal sacs ex-
panding to obliterate the outflow tract of right ven-
tricle (Fig. 1). After aneurysm resection had been
completed, the size of the defect was measured,
showing an average 0.55 ± 0.1 cm (range 0.3–1.4).

Twenty-four patients had AVP, and the aortic
valve was repaired in 13 in whom the regurgitation
was mild to moderate. The plication stitch was
placed at a single end of an aortic cusp in eight
patients and at two ends in five patients. In two pa-
tients with severe regurgitation aortic valve replace-
ment required. Postoperatively, there was no
evidence of aortic valve regurgitation in 11 patients,
and there were signs of only mild aortic valve regur-
gitation in two who had aortic valvuloplasty with
two plication stitches.

There was no operative death. The permanent
close atrioventricular heart block occurred in
one patient. Postoperative follow-up periods ranged
from 3 months to 15 years (average 5.6 ± 3.0 years).
All patients were controlled for recurrence of aneu-
rysms, tricuspid insufficiency and aortic regurgitation
by echocardiographic examinations. There were no
late deaths. Two patients underwent reoperation for
residual or recurrent shunt, two months and four
years postoperatively. In follow-up, new occurrences
of aortic regurgitation and increases of the persistent
regurgitation were not seen. We have not encoun-
tered any significant dysfunction of the tricuspid
valve resulting from incision and repair of the aneu-
rysma sac by postoperative Doppler echocardiogra-
phy examinations. There were no instances of late
endocarditis. All the patients in this study belong to
class I according to the functional classification of
the New York Heart Association.

4. Discussion

In our series, AMS was associated with severe com-
plexations. Aneurysm formation functionally reduced
the VSD size, but had the potential consequence of
promoting tricuspid insufficiency, aortic valve prolapse,
right ventricle outflow tract obstruction, rupture of the
aneurysm, and bacterial endocarditis.

Obstruction of the right ventricular outflow tract by
an aneurysm of the membranous septum has been
described [6,15,16]. In this study, in patients with VSDs
associated with an infundibular obstruction; defects
were moderate to large and gradients usually were
under 25 mmHg. However in two patients with AMS
with small shunt, the aneurysm itself made the gradient.
The LV-RA shunt associated with perimembranous
VSD is believed to be a result of anomalies of the
tricuspid valve. This shunt has also been described in
patients with an AMS associated with a VSD [14,17].
We have noted the presence of a systolic jet entering the
right atrium in some patients with an AMS. Changes in
the anatomy of the tricuspid valve relating to the
development of the AMS may facilitate the appearance
of a LV-RA shunt and tricuspid regurgitation.

An important finding of our study was the high
number of cases with aortic regurgitation (15/51,
29.4%). Aortic regurgitation has been reported by some
authors to occur between 5.5 and 18% of children or
adult patients with unoperated VSD [1,4,9,13]. In pa-
tients with AMS, Freedom and co-workers reported
that aortic valve prolapse developed in two of 32
patients (6.25%) [6]. The size of the VSD may be an
important factor for aortic valve prolapse and regur-
gitation. With time, the size of the defect in patients with
AMS is becoming smaller but the small size of the
defect produces the rapid blood flow velocities needed
to pull the aortic cusp into the ventricular septal defect,
which will eventually cause aortic valve prolapse and
regurgitation.

The reported incidence of bacterial endocarditis in
VSD has varied from 1/200 to 1/1200 patient-years,
depending on the patient population studied and the
method by which the information on the incidences was
tabulated [7,8]. Otterstad and colleagues reviewed 109
adult patients with VSD, and non operated patients
had 15% incidence of bacterial endocarditis [13]. In
the present series, the incidence of bacterial endocarditis
was 5.42% in adult patients with isolated perimem-
bra nous VSD that their hemodynamical size was usually
mild. Bacterial endocarditis has been documented as a
complication of AMS in several series [6,15]. In our
series, bacterial endocarditis was far more common in
patients with AMS than isolated VSD; the disease
occurred about three times as often in the patients with
AMS. LV-RA shunt which appears at a fairly high rate
after aneurysmal formation has a high risk of bacterial
endocarditis. The increased incidence of bacterial endocarditis may also be explained by fibrous patch formation. High-velocity jet stream created by blood passing a narrowed orifice of the aneurysm of membranous septum with a significant pressure gradient may allow platelets and fibrin to adhere to the partially denuded surface of roughened endothelium, creating a sterile thrombus that may entrap the microorganisms [5].

In our experience, direct suture closure of the VSD with aneurysm of the membranous septum is unsatisfactory. Residual communication and/or recurrence of the aneurysm were found postoperatively in two of nine direct suture closure in our series. The thick fibrous aneurysm tissue surrounding the defect may seem to be adequate for use as a buttress, but there may be multiple fenestration in the base of aneurysm and unless the aneurysm is excised completely, it may be overlooked. Also, the portion in the base of the aneurysm may weaken and cause further enlargement due to the effect of left ventricle pressure. In addition, the direct suture closure of aneurysm may cause distortion of tricuspid leaflets. Our current policy is to excise the aneurysm completely and close the produced true or actual septal leaflets. Our current policy is to excise the aneurysm and close the produced true or actual septal leaflets. Our current policy is to excise the aneurysm completely and close the produced true or actual septal leaflets.

Aneurysm formation may be an important mechanism in reducing the functional size of an associated membranous VSD, otherwise the aneurysm itself is associated with some severe complications which may be seen over an extended period in adulthood. In addition, anatomic closure of the VSD was not observed in all series concerning AMS [3,6,11,14,15]. Therefore we recommend that AMS should be operated on during childhood in order to prevent further enlargement and consequent complications even if there are no cardiac symptoms.

References