Echocardiography in Discrete Subaortic Stenosis

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SUMMARY Differentiating discrete subaortic stenosis (DSAS) from aortic valve stenosis (AVS) may be difficult, even at cardiac catheterization. Twenty-two patients with DSAS were studied echocardiographically and compared with 41 patients with AVS and 234 normal subjects. A ratio of left ventricular outflow tract to aortic root (LVOT:AO) < 0.80 determined LVOT narrowing in 19 of 22 DSAS patients. The aortic valve echogram was abnormal in 19 of 20 patients with DSAS and complete aortic valve echograms. Abnormalities included marked fluttering and early systolic partial closure ranging from 13–88% of total aortic valve systolic excursion. AVS patients did not have comparable changes. Neither LVOT narrowing nor degree of aortic valve partial closure (AVPC) correlated with pressure gradients determined by catheterization. A subaortic membrane was visualized in three patients. Asymmetric septal hypertrophy was noted in five patients with DSAS.

DSAS is characterized by LVOT narrowing, early systolic AVPC, marked aortic valve flutter, and, in some cases, by a subaortic membrane. Cardiac catheterization is necessary to quantitate the degree of LVOT obstruction.

ALTHOUGH DISCRETE SUBAORTIC STENOsis (DSAS) is less common than aortic valve stenosis (AVS), it is important in the differential diagnosis of left ventricular outflow tract (LVOT) obstruction and has unique surgical and prognostic implications. DSAS may clinically simulate AVS and a membrane or a fibromuscular structure located a few millimeters below the aortic valve may be difficult to identify, even at cardiac catheterization. Previous investigators have reported several echocardiographic signs which should differentiate DSAS from AVS.

In patients with DSAS we used echocardiography to assess the accuracy of echocardiographic differentiation of DSAS from AVS, to evaluate possible non-invasive prediction of outflow gradient or severity of stenosis, and to describe echocardiographic criteria for diagnosis of LVOT narrowing suggestive of DSAS.

Materials and Methods

Three groups of patients were studied echocardiographically and analyzed retrospectively. Group 1, a control group, consisted of 234 patients who had no clinical or cardiac catheterization evidence of asymmetric septal hypertrophy, AVS, or DSAS; patients with asymmetric septal hypertrophy demonstrated echocardiographically were excluded. These patients weighed from 11–103 kg. They were selected from an alphabetical analysis of echocardiographic files. To be included in the study, the aortic root, mitral valve, left interventricular septal surface, and left ventricular posterior wall echogram had to be clearly recorded and easily measurable throughout cardiac cycles on a continuous aortic-to-left-ventricular sweep. Of 234 aortic valve echograms, 182 (78%) had both anterior and posterior aortic leaflets recorded throughout systole and underwent detailed analysis. Group 2 consisted of 41 patients (weight range 11–103 kg) with AVS proven by cardiac catheterization. All patients had complete LVOT echograms, and the aortic valve was adequate for analysis in 32 (78%). Group 3 consisted of 22 patients (17 males and five females, 5–65 years old) with DSAS who were studied by echocardiography and cardiac catheterization.

Based on the clinical examination alone, DSAS was strongly suspected in four of 22 (18%) patients. Fifteen of 22 patients had echocardiograms before cardiac catheterization or surgery. A firm echocardiographic diagnosis was made prospectively in nine of these 15 patients (56%). The diagnosis was not made on initial cardiac catheterization in seven of 22 patients (32%). These patients had no echocardiographic studies before catheterization. Six of these seven later had echocardiograms (either before repeat catheterization or surgery) which suggested DSAS, and a definite echocardiographic diagnosis was made in five. Catheterization or surgery confirmed the diagnosis in all seven patients. Twelve patients with DSAS underwent surgery; 10 survived (83%). Eight of 10 patients underwent postoperative echocardiographic examination and four were recatheterized.

The echocardiographic studies were performed with an Ekoline 20 ultrasonoscope coupled to a Cambridge, Honeywell, or Irex recorder, using a 2.25 MHz transducer focused at 5 or 7.5 cm. A Unirad ultrasonoscope and Honeywell Visicorder were used for one patient. The transducer was positioned at the left sternal border in the interspace, which allowed us to record the mitral valve with the transducer perpen-
any other occurrence of the term 'LVOT' is the left ventricular outflow tract. LVOT to aortic valve ratio was considered abnormal if the ratio was less then the weight limits established in Group 1: for example, LVOT < 1.4 cm in a patient weighing 11–23 kg, or an LVOT:AO ratio < 0.80 with normal aortic size.16 The LVOT dimension and LVOT:AO ratio were not decreased in any patient with AVS. In Group 3 the LVOT:AO ratio was decreased in 19 of 22 patients (fig. 4) while the LVOT measurement was below the weight limits in 11 patients (table 1). The LVOT:AO ratio was decreased

mitral leaflet to the left septal surface on the cycle showing motion characteristic of the mitral valve leaflet.16 Multiple straight-line sweeps along the long axis of the LVOT were attempted. To minimize artificial narrowing of the LVOT, we selected the sweep with the greatest LVOT dimension in each patient. Within the selected sweep, maximal narrowing was identified by selecting the cardiac cycle with the smallest LVOT dimension (figs. 1 and 2). We used the LVOT:AO ratio to evaluate the severity of LVOT narrowing.

Maximum separation of the anterior and posterior aortic valve cusps was measured at the opening and immediately after aortic valve partial closure on the cycle that showed the greatest amount of partial closure. We calculated the percentage of partial closure by dividing the initial maximum aortic valve separation (AVmax) minus the minimum aortic valve separation (AVmin) by the AVmax and multiplying by 100 (fig. 3). Left ventricular catheterization was performed using retrograde arterial or transseptal techniques. LVOT gradients were measured by pullback recording using, in most cases, an end-hole catheter or simultaneous left ventricular and aortic recording.

Results

The values measured for AO and LVOT and the resulting LVOT:AO ratios are summarized in table 1. The normal range of the LVOT:AO ratio was 0.80–1.50 (fig. 4). The LVOT was considered narrowed if it was less than the weight limits established in Group 1: for example, LVOT < 1.4 cm in a patient weighing 11–23 kg, or an LVOT:AO ratio < 0.80 with normal aortic size.16 The LVOT dimension and LVOT:AO ratio were not decreased in any patient with AVS. In Group 3 the LVOT:AO ratio was decreased in 19 of 22 patients (fig. 4) while the LVOT measurement was below the weight limits in 11 patients (table 1). The LVOT:AO ratio was decreased

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in the two patients with inadequate aortic valve echo-
grams and the one patient with a normal aortic valve
study. The severity of LVOT narrowing (LVOT: AO
ratio) did not correlate significantly with the LVOT
systolic gradient obtained at catheterization.

Nine patients in the control group had systolic aor-
tic valve partial closure ranging from 10–22%, while no
patients with AVS had partial closure. Nineteen of 20
aortic valves recorded in patients with DSAS had par-
tial closure immediately after opening (fig. 5). The
partial closure ranged from 13–88% (p < 0.01; DSAS
vs control), but the partial closure percentage did not
correlate significantly with the severity of obstruction
as judged by LVOT systolic pressure gradient.

Exaggerated systolic fluttering of the aortic valve
cups was present in 19 of the 20 DSAS patients with
adequate records (fig. 5). Twenty-eight patients in the
control group had high frequency aortic valve systolic
fluttering (fig. 6A). However, no patient in the control
group had the coarse, low frequency component seen
in many patients with DSAS (fig. 5). No patient with
valvular stenosis had this degree of high or low fre-
quency systolic valve fluttering.

Echocardiographic criteria alone did not con-
sistently predict whether the DSAS was a membrane
or a fibromuscular segment or a combination of the
two. In patients with fibromuscular segments, es-
timates of segment length did not correlate well with
anatomic findings because of beam angulation and

TABLE 1. Summary of Echocardiographic Findings

<table>
<thead>
<tr>
<th>Weight (kg)</th>
<th>N</th>
<th>AO (cm)</th>
<th>LVOT (cm)</th>
<th>LVOT:AO</th>
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<tr>
<td></td>
<td></td>
<td>Range</td>
<td>Mean</td>
<td>Range</td>
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<tr>
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<td>47</td>
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<td>1.71</td>
<td>1.4–2.4</td>
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<td>24–33</td>
<td>29</td>
<td>1.6–2.5</td>
<td>1.94</td>
<td>1.6–2.5</td>
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<tr>
<td>34–45</td>
<td>22</td>
<td>1.9–3.0</td>
<td>2.23</td>
<td>1.9–3.0</td>
</tr>
<tr>
<td>46–57</td>
<td>49</td>
<td>1.9–2.9</td>
<td>2.32</td>
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<tr>
<td>&gt;58</td>
<td>87</td>
<td>2.0–3.6</td>
<td>2.56</td>
<td>1.9–3.3</td>
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Summary of echocardiograms in aortic stenosis (group 2)

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<th>Range</th>
<th>Mean</th>
<th>Range</th>
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<th>Range</th>
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<td>1.86</td>
<td>1.06–1.11</td>
<td>1.09</td>
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<td>24–33</td>
<td>8</td>
<td>1.6–2.5</td>
<td>1.95</td>
<td>2.0–2.5</td>
<td>2.20</td>
<td>1.00–1.37</td>
<td>1.15</td>
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<td>34–45</td>
<td>6</td>
<td>2.1–2.7</td>
<td>2.33</td>
<td>2.1–2.3</td>
<td>2.20</td>
<td>0.85–1.00</td>
<td>1.20</td>
</tr>
<tr>
<td>46–57</td>
<td>6</td>
<td>1.9–2.6</td>
<td>2.26</td>
<td>2.3–3.2</td>
<td>2.70</td>
<td>1.00–1.39</td>
<td>1.20</td>
</tr>
<tr>
<td>&gt;58</td>
<td>17</td>
<td>2.0–3.6</td>
<td>2.91</td>
<td>2.2–3.6</td>
<td>2.95</td>
<td>0.93–1.29</td>
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Summary of echocardiograms in subaortic stenosis (group 3)

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<th>Range</th>
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<tbody>
<tr>
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<td>1.8–3.6</td>
<td>2.53</td>
<td>0.8–3.6</td>
<td>1.51</td>
<td>0.38–1.00</td>
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<td>2.90</td>
<td>0.9–1.9</td>
<td>1.33</td>
<td>0.30–0.56</td>
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<tr>
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<td>0.46–0.64</td>
<td>0.55</td>
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<tr>
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<td>3.0–3.1</td>
<td>3.03</td>
<td>0.8–2.2</td>
<td>1.60</td>
<td>0.26–0.70</td>
<td>0.53</td>
</tr>
<tr>
<td>&gt;58</td>
<td>6</td>
<td>2.6–3.6</td>
<td>3.27</td>
<td>1.0–3.6</td>
<td>1.88</td>
<td>0.37–1.00</td>
<td>0.56</td>
</tr>
</tbody>
</table>

Abbreviations: N = number of patients; AO = aortic root; LVOT = left ventricular outflow tract.
FIGURE 4. Left ventricular outflow tract to aortic root ratio (LVOT:AO). Individual data points are indicated for aortic valve stenosis, group 2 (AVS) and discrete aortic valve stenosis, group 3 (DSAS). Mean and SD are indicated by horizontal line and brackets, respectively. The area shaded with diagonal lines indicates the range for the control group (NORM) with mean and SD indicated by the horizontal line and brackets, respectively. The number of data points (patients) equals 234.

variability in LVOT sweep time. A subaortic membrane was recorded in three patients previously reported. The membrane could be differentiated from systolic anterior mitral valve motion because it was recorded anterior to the anterior mitral leaflet throughout the cardiac cycle. The mitral valve echograms of two patients showed total diastolic septal abutment and a consequently flat diastolic E-F slope. Catheterization showed only a subaortic membrane. One additional patient whose echogram suggested only a membrane was found at surgery to have a membrane with a fibromuscular base.

Twelve of 20 DSAS patients who had aortic root

FIGURE 5. Aortic valve echograms from three patients with discrete subaortic stenosis (DSAS). Note aortic valve partial closure in early systole (black arrows). Also note the coarse, low-frequency flutter of the anterior aortic valve leaflet in the right panel and the high frequency valve flutter in all three panels. ECG = electrocardiogram; LA = left atrium.
angiography had aortic regurgitation, but on the echocardiogram, only five had high frequency diastolic flutter of the mitral valve suggesting aortic regurgitation.

Five patients with DSAS had asymmetric septal hypertrophy preoperatively (IVS:PW range 2.8–1.3). Two patients with IVS:PW ratios of 1.6 and 1.4 did not have follow-up studies. In three patients who had follow-up studies, preoperative IVS:PW ratios were 2.8, 1.4 and 1.3. The postoperative ratios in these patients were 1.7, 1.1 and 1.0, respectively.

Postoperative echocardiograms were done on eight of 10 patients who survived surgery (fig. 7). The LVOT:AO ratio increased in four patients, decreased in one patient and was unchanged in three patients. The LVOT measurement increased in four, decreased in two and was unchanged in two. The aortic valve partial closure percentage increased in seven patients and was unchanged in one.

Discussion

Prior reports have described echocardiographic features of DSAS.5–11 Davis et al.6 reported systolic aortic valve partial closure and marked fluttering of aortic valve cusps in three patients with DSAS. Popp et al.4 reported narrowing of the LVOT in three patients with DSAS. Direct recording of the subaortic membrane has been previously reported.6, 7–11 However, Goldberg et al.17 were unable to confirm these findings in three patients with DSAS.

The aortic valve is often difficult to record in patients with DSAS, but with persistent examination by experienced operators, good aortic valve echocardiograms were obtained in 20 of 22 patients in this series. Aortic valve systolic fluttering is not specific for DSAS, as it occurs in normal patients and a variety of abnormal conditions. However, its occurrence is rare in valve stenosis.

Partial valve closure may also occur in other conditions including ventricular septal defect and idiopathic hypertrophic subaortic stenosis (IHSS) (fig. 6C).18 The partial closure in IHSS is usually more midsystolic than that seen in our patients with DSAS. Although cases of isolated ventricular septal defect with aortic valve partial closure have been reported, the incidence of associated ventricular septal defect and DSAS suggests that the latter be sought in the presence of echocardiographic partial valve closure.6, 19 In our series, one patient had a septal defect associated with DSAS.

The aortic valve echocardiographic abnormalities do not correlate well with left ventricular aortic pressure gradients. Application of fluid dynamics principles provides a theoretical but plausible explanation. As the blood passes through the stenotic subaortic area a jet stream is formed and Bernoulli’s principle applies. The initial impact of the jet opens wide the valve, just as in the normal heart. However, as the jet of blood passes through the flexible valve cusps, the low pressure area bordering the jet lets the cusps move toward each other and the continuing flow causes in-
stability and a fluttering motion (fig. 8). When viewed echocardiographically, the motion (partial closure) may be abrupt, followed by lesser vibrations in the valve, or may be a series of coarse oscillations (fig. 5). Frequently, the effect on the two cusps visualized is asymmetric. This phenomenon may result from the jet being directed toward a particular area of the root by the distorted outflow tract. The Coanda effect, a fluid dynamics principle which has been applied to other asymmetric flow phenomena,20, 21 may also affect the course of the subaortic jet stream. These theories suggest that the configuration and orientation of the jet stream as well as the proximity of the point of obstruction to the plane of the aortic valve may determine the echocardiographic configuration of partial closure in DSAS relatively independent of the degree of obstruction.

Mild partial closure and systolic fluttering in normal patients are probably created by the same basic dynamic changes as they are in patients with DSAS.

**Figure 7.** Echocardiographic sweep in a patient after surgery for DSAS. Left ventricular outflow tract to aortic root (LVOT:AO) ratio continues to be abnormal (0.7). Multiple sites of measurement are designated by vertical arrows. Open arrows identify sites selected for measurement. LA = left atrium; MV = mitral valve; S = septum.

**Figure 8.** Diagrammatic representation of aortic valve partial closure during systole. A) During the initial phase of left ventricular ejection the aortic valve opens widely as a result of left ventricular output and the subaortic jet stream. B) As the jet passes through the flexible valve leaflets, low pressure areas bordering the jet stream create a pressure differential favoring leaflet closure. C) As flow continues, turbulence and instability develop resulting in lesser oscillations of valve leaflets. ECG, echocardiographic trace, and diagrammatic representation of subaortic stenosis are presented. Dotted line in echo panel represents typical systolic configuration of a normal aortic valve. ECG = electrocardiogram; ECHO = echocardiogram.
The effect is much less pronounced because the jet of blood is broader and its velocity less. The damping effect of the thick, noncompliant leaflet(s) in the patient with AVS would make the valve unresponsive to these dynamic pressure changes and explain the absence of flutter or partial closure.

Gramiak and Shah found LVOT narrowing (defined as less than 2.0 cm) in normal adults and patients with IHSS, as well as several other lesions clinically distinct from aortic stenosis. We defined LVOT narrowing as an LVOT:AO ratio < 0.80 and we have seen this degree of narrowing in patients with asymmetric septal hypertrophy. Nineteen of 22 patients with DSAS had LVOT narrowing according to this criterion, but the ratio did not correlate significantly with the severity of obstruction. The LVOT:AO ratio did allow identification of the three patients without aortic valve partial closure. Careful measurement of the aortic diameter is necessary because aortic dilatation invalidates the LVOT:AO ratio.

Transsecting the true LVOT diameter with the echobeam can be difficult and is a potential source of error. However, using multiple sweeps minimizes the problem of sweeping off center through the obstructed outflow tract and has proved reliable in our series. Cross-sectional ultrasonography provides a more complete view of the distorted LVOT. However, until cross-sectional instruments become more widely available and resolution is improved, the noninvasive diagnosis of many patients with DSAS will be made with one-dimensional echocardiography.

The poor correlation between the left ventricular outflow gradient and the observed narrowing is not entirely unexpected. The subaortic orifice would have to form a regular geometric shape, such as a cylinder of fixed length, for the one-dimensional M-mode examination to provide a consistently accurate estimate of the outflow area. The more distorted the outflow tract, the greater will be the potential error in the estimate.

The presence of a membrane demonstrated by echocardiography was confirmed by surgery or angiography in three of the 22 DSAS patients. However, a membrane was diagnosed by angiography or surgery and was not demonstrated by echocardiography in eight patients. With one exception, the patients with a decrease in the LVOT:AO ratio had anatomic LVOT narrowing with or without a discrete membrane. We could not record the membrane consistently, probably because it was oriented parallel to the sound beam.

A subaortic membrane was recorded in several previously reported patients. The motion of the membrane was similar in all cases. Echocardiographic differentiation of a subaortic membrane from systolic anterior motion of the mitral valve is possible because the membrane is recorded anterior to the mitral valve throughout the cardiac cycle. In addition, the membrane and anterior mitral leaflet are recorded simultaneously, but clearly separate from each other in systole and late diastole.

Outflow tract narrowing and systolic aortic valve partial closure were not observed in patients with valvular stenosis. Conversely, we have not yet observed DSAS patients with the severely thickened or calcified leaflets as seen in valvular stenosis (fig. 6B). IHSS with aortic valve partial closure can be identified by an adynamic septum, asymmetric septal hypertrophy, the presence of typical systolic anterior motion, and anterior displacement of the mitral valve.

In conclusion, clinical findings of LVOT obstruction with marked systolic aortic valve flutter and early systolic aortic valve partial closure rule out isolated valvular stenosis. Very early partial closure and valve flutter are typical of DSAS and were commonly found in our series. LVOT:AO ratios < 0.80 are found in a high proportion of DSAS patients, but the subaortic membrane per se may not be recorded, even when present. The echocardiographic features of DSAS are clear enough and specific enough to be used in assessing patients with clinical signs of LVOT obstruction.

References

Subxiphoid Cross-Sectional Echocardiography in Infants and Children with Congenital Heart Disease

LOTHAR W. LANGE, M.D., DAVID J. SAHN, M.D., HUGH D. ALLEN, M.D., AND STANLEY J. GOLDBERG, M.D.

SUMMARY We developed anatomically related cross-sectional echocardiographic views to image the heart from the subxiphoid area in 100 children and infants with various forms of congenital heart disease studied prospectively before cardiac catheterization. Wide-angle cross-sectional views were achieved using a mechanical sector scanner in a scan plane oriented parallel to a line between the patient’s shoulders (a coronal plane) showing the equivalent anatomy of an anteroposterior angiogram. The subxiphoid technique appeared to be better than chest wall-based imaging in demonstrating obstructive lesions in the proximal portion of the right ventricular outflow tract (19 patients), i.e., the subpulmonic area, which is often at the narrow edge of the sector and behind the transducer artifact in chest wall studies. The subxiphoid technique was also useful for imaging the interatrial and interventricular septae; in subxiphoid views, as opposed to four-chamber apical views, there was significantly less false septal dropout and atrial septal defects (19 patients) as well as ventricular septal aneurysms (seven patients) were easily imaged. Finally, the subxiphoid orientation provided more adequate imaging in patients with discrete diaphragmatic subaortic stenosis (four patients), even when the diaphragm was just beneath the aortic valve. Subxiphoid cross-sectional echocardiography is an easily understood anatomical format for imaging cross-sectional anatomy in congenital heart disease and is a valuable adjunct to cross-sectional echocardiography from the chest wall.

CROSS-SECTIONAL ECHOCARDIOGRAPHY allows noninvasive assessment of cardiac spatial anatomy which is especially useful when evaluating children with congenital heart disease.1-3 Moreover, anatomic details can often be demonstrated more accurately with this method than with single-crystal M-mode imaging. The usual planes of examination require placement of the transducer on the chest wall, which is a limiting factor in patients with chest wall deformities or lung disease. The subxiphoid M-mode technique was first introduced by Chang et al.4 as an alternative method for visualizing the left ventricle and left ventricular outflow tract. In this study, we have verified and evaluated cross-sectional planes for cardiac examinations from a subxiphoid transducer position, as initially described by us and others.5, 4, 5

The subxiphoid examination is easy to perform and presents easily understood coronal plane anatomy. We studied 100 infants and children with congenital heart disease to determine the usefulness of cross-sectional echocardiography from the subxiphoid position for imaging the right and left atrial cavities, right and left ventricular cavities, the atrial and ventricular septae, and the right and left ventricular outflow tracts.

Methods

Patients (table 1)

We performed a preliminary study of 18 children who did not have cardiovascular disease (ages 2–5.5 years) to identify the structures and cavities which could be imaged from subxiphoid placement of the transducer. These studies were performed after obtaining informed parental consent as part of a Human Subjects Approved Research protocol. Right-sided structures were verified by peripheral venous echo contrast injections6 of 3–5 ml of normal saline solution. Once the identification process was completed, the subxiphoid view was included as routine in
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