Open aortic valvotomy for congenital aortic stenosis

Late results

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SUMMARY Forty-nine consecutive patients, aged 2 to 28 years, were followed after open aortic valvotomy. Three late deaths occurred in relation to reoperation.

Seventeen reoperations were performed 2 to 14 years after valvotomy for severe stenosis in 12 patients, aortic regurgitation in three patients, and aortic stenosis and regurgitation in two patients. Among the 12 patients who required reoperation for severe obstruction, five aged over 19 years had calcified valves with normal aortic roots and valve replacement was simple. Seven had tunnel obstruction with a hypoplastic aortic root, constituting a difficult surgical problem, and necessitating total aortic root replacement in four.

The postoperative course after simple aortic valvotomy is determined by several factors; the basic pathological form of the obstruction is the most important. Those who present in the first decade with lumpy valves and small aortic roots tend to form a diffuse tunnel obstruction when residual stenosis remains after valvotomy; older patients with pliable domed valves slowly develop calcified cusps and present less problems as the aortic root is usually a good size.

Although aortic valvotomy offers good early results with a low mortality, it should be regarded as palliative as all patients will ultimately require reoperation. Younger patients with lumpy valves and a small aortic root have more problems and may require different initial management.

Open aortic valvotomy for relief of congenital aortic valve obstruction in children and adolescents is associated with a low mortality and results are usually reported to be good or excellent.1–6 Since the abnormal aortic valve is ultimately destined to calcify, there must be concern about the long-term future of these patients, who eventually will require aortic valve replacement.7–9 This study was undertaken to review the fate of these patients and to see if any new or unexpected problems were encountered.

Subjects and methods

Data on fifty-two patients with congenital aortic valve stenosis between 1961 and 1978 are presented. The age distribution of the patients at the time of operation is shown (Fig. 1)

Symptoms were present in 24 patients; dyspnoea

Fig. 1 Ages of 52 patients at first open aortic valvotomy. The peak incidence occurred at 8 to 9 years and around puberty, between 12 and 15 years.
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valve was 50 to 160 mmHg in 43 patients, exceeding 100 mmHg in 14 patients. Two patients operated on with gradients below 50 mmHg across the valve had unusual septal hypertrophy which was disproportionate to the degree of valvar stenosis; in these patients it was considered necessary to remove the valvar obstruction as a possible stimulus to excessive myocardial dysplasia.10 The aortic valve was not crossed in three patients; the aortogram confirmed the narrow orifice of the valve and the electrocardiogram showed grade 2 left ventricular hypertrophy in one patient and grade 3 in the other two. Six patients had mild aortic regurgitation.

Table 1 Associated congenital cardiovascular anomalies in 16 of 52 patients who had open aortic valvotomy

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coarctation</td>
<td>6</td>
</tr>
<tr>
<td>Persistent ductus arteriosus</td>
<td>4</td>
</tr>
<tr>
<td>Fixed subvalvar stenosis</td>
<td>3</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>1</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>1</td>
</tr>
<tr>
<td>Mitral regurgitation</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary valve stenosis</td>
<td>1   (Rubella)</td>
</tr>
</tbody>
</table>

The associated congenital cardiovascular anomalies present in 16 patients are summarised (Table 1). Six patients had had previous surgery: three had had resection of coarctation, one had had resection of coarctation and ligation of persistent ductus arteriosus, one had had ligation of a persistent ductus arteriosus, and one a pulmonary valvotomy and ligation of a persistent ductus arteriosus.

Open aortic valvotomy was performed using cardiopulmonary bypass with normothermia or moderate hypothermia. Myocardial protection with infusion of Ringers potassium solution has been used in the National Heart Hospital since 1978. After opening the ascending aorta to expose the abnormal aortic valve, the valvotomy consisted of opening the two lateral commissures but never the anterior fused raphe. Often, despite high gradients, there was little the surgeon could do to open the valve without increasing aortic regurgitation, particularly with true bicuspid valves. In 15 patients a subvalvar myotomy was also performed because of severe subvalvar septal hypertrophy. Two patients died in the perioperative period, one of whom had a thick lumpy valve and a small aortic root, and one patient required an aortic valve replacement during the immediate postoperative period. The other 49 patients who left hospital have been followed up for two to 18 years. No patient has been lost to follow-up. All had clinical, electrocardiographic, and radiological assessment. Thirty-seven had repeat cardiac catheterisation, aortography, and left ventricular angiography.

Results

ANATOMIC FINDINGS

In 42 patients the valve was described as "bicuspis". On reviewing the operative findings, the valve was truly bicuspis with two symmetrical or asymmetrical cusps in only 16 patients. In the other 26 the anterior cusp with the two coronary ostia was slightly larger than the posterior one and contained a rudimentary central raphe which was never opened. Truly tricuspid aortic valves were found in seven patients; in the other three patients, aged 3, 9, and 19 years, it was
not possible to recognise the commissures.

Slight calcification of the valve was found in five patients, all of whom were older than 18 years.

In 14 patients the valve was thick and lumpy with nodular excrescences, sometimes resembling the cartilage of the ears (Fig. 2); 11 of these valves were bicuspid. The age of these 14 patients varied from 3 to 28 years (mean 10 years). In the other 38 patients the valves were thin and pliable.

Varying degrees of aortic root dilatation were present in all but 12 of the patients where the ascending aorta was narrow and small, as shown by angiography and confirmed at surgery. A supravalvar waist was present in the hypoplastic aortic root in six of these 12. Six of the patients with thick lumpy valves also presented with a narrow aortic root.

In 15 patients in whom myotomy and wedge resection were carried out the left ventricle was extremely hypertrophied particularly in the area of the septum. In five the angiographic findings suggested an unusually thick septum and irregular muscular thickening resembling appearances characteristic of hypertrophic cardiomyopathy. There were no correlations between the degree of hypertrophy of the left ventricle, the gradient found at cardiac catheterisation, and/or the anatomy of the aortic valve or the outflow tract.

REOPERATION

Seventeen of the 49 patients (34.7%) had a second operation on the aortic valve two to 14 years after the aortic valvotomy (Fig. 3); three patients died at reoperation. The principal indication for reoperation was severe obstruction (12), aortic regurgitation (three), or a combination of both lesions in two (Table 2). The peak systolic gradient was measured before reoperation in 14 patients and ranged from 30 to 125 mmHg (mean 79 mmHg). In most patients the resting gradient before reoperation was not as high as before the first operation (Fig. 4). The electrocardiogram became worse in seven patients and remained unchanged in the other 10. Five of the 12 patients with severe aortic stenosis had severe calcification of the valve, one of whom died after reoperation. These were the oldest patients.

Seven patients had a long “tunnel” obstruction with a hypoplastic aortic root and ring. Six had thick, lumpy valves, with nodular excrescences in three. Supravalvar waisting and subvalvular muscular hypertrophy were present in all, thus constituting a diffuse obstruction of the whole left ventricular outflow tract (Fig. 5). To relieve this diffuse obstruction four patients had total aortic valve and root replacement with a fresh, antibiotic sterilised aortic homograft with reimplantation of the coronary arteries,11 12 two had their valve replaced with enlargement of the aortic root with a Dacron patch; one of these patients required further operation seven years later and died during this. The seventh patient had excision of a fibrous stricture from beneath the valve which had been removed two years earlier at the time of aortic valvotomy. The valve at this time was found to be competent and not stenotic and the aortic root that had been enlarged at the time of valvotomy with a Dacron patch was found to be normal.

Three patients who had dominant aortic regurgitation had a second operation five to eight years after valvotomy. One patient, with a hypoplastic aortic root, developed severe aortic regurgitation six years after valvotomy because of detachment of one of the lumpy cusps of a bicuspid valve; he died at reoperation. In the other two, mild aortic regurgitation had been present since the valvotomy but did not become important until two and five years respectively after the first operation. At that time the valve was found to be rigid and lumpy with excrescences of cartilaginous substance in one patient and thick in the other.

Two patients presented with aortic stenosis and regurgitation requiring reoperation six and eight years after valvotomy. Both had had thick, lumpy valves with nodular excrescences which were excised. At first, aortic regurgitation was mild but it progressed suddenly after five to seven years and both were found to have holes in the left coronary cusp which had developed where the excrescences had been excised, leaving holes which had been repaired with prolene sutures. One had aortic root replacement for a small aortic root and the other had an aortic homograft valve replacement which required removal four years.
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Table 2 Summary of data on patients requiring reoperation, valve used for replacement, and subsequent outcome

<table>
<thead>
<tr>
<th>Age at 1st opn</th>
<th>Age at reopn</th>
<th>Indication reopn</th>
<th>Findings at 2nd opn</th>
<th>Type of opn</th>
<th>Valve used for replacement</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>17</td>
<td>23</td>
<td>AS</td>
<td>Calcific aortic valve</td>
<td>AVR + myotomy</td>
<td>Pulmonary autograft</td>
<td>Well</td>
</tr>
<tr>
<td>19</td>
<td>25</td>
<td>AR</td>
<td>Lumpy aortic valve + small aortic root</td>
<td>AVR + myotomy + enlarged aortic root</td>
<td>Pulmonary autograft</td>
<td>Died at 3rd operation</td>
</tr>
<tr>
<td>11</td>
<td>16</td>
<td>AR</td>
<td>Thick fibrotic aortic valve</td>
<td>AVR</td>
<td>Homograft</td>
<td>Well</td>
</tr>
<tr>
<td>6</td>
<td>14</td>
<td>AS</td>
<td>Thick aortic valve + small aortic root</td>
<td>AVR + root replacement + myotomy</td>
<td>Homograft aortic root + valve</td>
<td>Well</td>
</tr>
<tr>
<td>28</td>
<td>35</td>
<td>AS + AR</td>
<td>Calcific aortic valve</td>
<td>AVR</td>
<td>Homograft</td>
<td>Died</td>
</tr>
<tr>
<td>15</td>
<td>21</td>
<td>AS + AR</td>
<td>Lumpy aortic valve + hole cusp + small aortic root</td>
<td>AVR + root replacement</td>
<td>Homograft aortic root + valve</td>
<td>Well</td>
</tr>
<tr>
<td>6</td>
<td>14</td>
<td>AS + AR</td>
<td>Lumpy aortic valve + hole cusp</td>
<td>AVR</td>
<td>Homograft</td>
<td>4 y later ao root + valve repl.</td>
</tr>
<tr>
<td>27</td>
<td>35</td>
<td>AS</td>
<td>Calcific aortic valve</td>
<td>AVR</td>
<td>Homograft</td>
<td>Well</td>
</tr>
<tr>
<td>8</td>
<td>22</td>
<td>AS</td>
<td>Lumpy aortic valve + small aortic root</td>
<td>AVR + root replacement</td>
<td>Homograft aortic root + valve</td>
<td>Well</td>
</tr>
<tr>
<td>13</td>
<td>15</td>
<td>AS</td>
<td>Aortic valve thin, competent, + subaortic ring, + small aortic root</td>
<td>Relief of subaortic obstruction</td>
<td>Well</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>15</td>
<td>AR</td>
<td>Lumpy aortic valve, + detachment of cusp, + small aortic root</td>
<td>AVR + myotomy</td>
<td>Homograft</td>
<td>Died</td>
</tr>
<tr>
<td>3</td>
<td>8</td>
<td>AS</td>
<td>Lumpy aortic valve, + small aortic root</td>
<td>AVR + root replacement</td>
<td>Homograft aortic root + valve</td>
<td>Well</td>
</tr>
<tr>
<td>9</td>
<td>23</td>
<td>AS</td>
<td>Calcific aortic valve</td>
<td>AVR + myotomy</td>
<td>Homograft</td>
<td>Well</td>
</tr>
<tr>
<td>5</td>
<td>8</td>
<td>AS</td>
<td>Thick aortic valve, + small aortic root</td>
<td>AVR + root replacement + myotomy</td>
<td>Homograft aortic root + valve, + pacemaker</td>
<td>Well</td>
</tr>
<tr>
<td>9</td>
<td>17</td>
<td>AS</td>
<td>Thick, lumpy aortic valve</td>
<td>AVR</td>
<td>Homograft</td>
<td>Well</td>
</tr>
<tr>
<td>15</td>
<td>29</td>
<td>AS</td>
<td>Calcific aortic valve</td>
<td>AVR</td>
<td>Homograft</td>
<td>2 y later AVR</td>
</tr>
</tbody>
</table>

AVR, aortic valve replacement; opn, operation; AS aortic stenosis; AR, aortic regurgitation.

later because of calcification.

No patient had a history or any signs of infective endocarditis at reoperation.

One 10-year-old boy required mitral valve surgery five months after aortic valvotomy for iatrogenic mitral regurgitation. This was a patient with severe subvalvular hypertrophy who had had a deep myotomy and wedge resection of myocardium which had disturbed the submitral valvar mechanism leading to ruptured chordae with a flail anterior cusp.

STATE OF SURVIVORS

Thirty-two patients have not yet had a second operation during the two to 18 years of surveillance (mean 6·3 years). Twenty have had cardiac catheterisation during the follow-up period and two patients had catheterisation repeated five and seven years after the first study.

Fifteen patients had moderate residual lesions with a gradient of 30 to 70 mmHg (Table 3); only three of these patients have symptoms. In seven patients with
predominant stenosis, signs of obstruction were still present immediately after the valvotomy but they have remained relatively stable during the following years. The two who both had repeated catheterisation at 5 and 7 years after their first study did not show any changes in gradient. Two patients with progressive obstruction had thick lumpy valves and a small aortic root. Among the patients with moderate aortic regurgitation, a mild early diastolic murmur was heard immediately after the operation in only five. No patient had bacterial endocarditis during the period of observation. No correlation was found between the presence and the degree of aortic regurgitation and the anatomical condition of the valve (bicuspid or tricuspid, lumpy or pliable) and/or the characteristics of the aortic root.

Thirteen asymptomatic patients had aortic valve stenosis with a resting gradient below 30 mmHg and/or trivial aortic regurgitation, static for one to 11 years. Three of these had lumpy valves and a small aortic root, factors that seem to predispose to the development of “tunnel” obstruction when residual obstruction is left behind; the aortic root had been enlarged with a pericardial patch at the time of valvotomy in all these.

Four of the 49 patients had excellent results and three to four years after valvotomy retained only a short soft systolic murmur with a late click and a normal electrocardiogram. Severe calcification of the aortic valve developed gradually in five patients above the age of 19 years followed for six to 18 years, with moderate stenosis.

Twelve patients who presented at the time of the first operation with extreme left ventricular hypertrophy on angiography were also given isoprenaline at postoperative cardiac catheterisation; in nine (Fig. 6) the subvalvar gradient was trebled. It was decided to give propanolol to four of these patients in the hope of preventing further hypertrophy and subvalvar obstruction as the electrocardiogram continued to show grade 3 changes and angiography showed severe and irregular muscular hypertrophy.

Fig. 7 shows the actuarial curve of survival following aortic valvotomy. After 18 years of follow-up 80% of patients were alive, with 98% survival in the first seven years. The proportion of patients alive with their own aortic valve (without reoperation) at 20 years, however, was only 14%, which means that if the standard error of 12% is taken into account, all patients will require reoperation within 20 years of follow-up.

Discussion

This group of patients is typical of congenital aortic valve stenosis which presents in childhood and adolescence. It excludes infants who present with critical aortic valve stenosis, however, whose problems are different since there is so much infarction (necrosis) in the left ventricle and the cusps are always poorly formed.

In most of the series of congenital aortic valve stenosis that we have reviewed3-13 there is no mention of the unusual form of the thick lumpy valve with a small aortic root. Some authors3 describe “some thick valves with thickening of the cusps particularly along the leading edges”. Other authors5 describe multiple levels of stenosis. In 1971 Somerville and Ross pointed out an unusual form of congenital aortic stenosis which may be part of a more diffuse abnormality. Fisher et al8 described three out of 45 patients with various forms of congenital aortic stenosis who presented with "a tunnel aortic stenosis" characterized by hypoplasia of the aortic valve ring and ventricular outflow tract, by thickened valve leaflet and by underdeveloped ascending aorta". They pointed out that this posed a difficult technical problem to relieve.

In the present series it is clear that the late mortality is related to reoperation, as in other series.17 A cause for concern is that the operative mortality for the first
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Fig. 5  Left ventricular angiograms from a patient with a tunnel obstruction who needed aortic valve and root replacement: small aorta with supravalvular waist, thick distorted aortic valve, and extreme hypertrophy of muscle in the left ventricle are present. The patient aged 13 years had the first aortic valvotomy at the age of 3 years.

Table 3  State of 32 patients who have had open aortic valvotomy and have not yet required reoperation

<table>
<thead>
<tr>
<th></th>
<th>Calcified aortic valve</th>
<th>Lumpy valve + small aortic root</th>
<th>Static since operation (follow-up years)</th>
<th>Progressive Electrocardiogram</th>
<th>Improved</th>
<th>Unchanged</th>
<th>Deteriorated</th>
</tr>
</thead>
<tbody>
<tr>
<td>Moderate aortic stenosis gradient 30–70</td>
<td>5</td>
<td>3</td>
<td>2</td>
<td>3 (4–13)</td>
<td>2 (2–11)</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>Aortic stenosis aortic regurgitation</td>
<td>10</td>
<td>2</td>
<td>4</td>
<td>6 (5–8)</td>
<td>4</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Mild aortic stenosis gradient &lt;30</td>
<td>13</td>
<td>3</td>
<td>13</td>
<td>7</td>
<td>5</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Excellent</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>32</td>
<td>5</td>
<td>5</td>
<td>24</td>
<td>8</td>
<td>15</td>
<td>13</td>
</tr>
</tbody>
</table>
Isoprenaline gradient in all nine patients.

The problem of progressive aortic regurgitation was small in this group, though other reports suggest it is the most common cause for reoperation being needed.\(^{15} 16\) Perhaps the careful techniques used here prevented important regurgitation but left more obstruction. Where myxomatous masses of nodular excrescences had been excised, the cusps appeared to weaken and later rupture, causing an acute increase in aortic regurgitation. Thus, this procedure should not be undertaken lightly even though it is simple. Patients with the small roots and lumpy valves where left with aortic regurgitation were easier to manage at reoperation as tunnel obstruction did not form.

Infected endocarditis was not responsible for any deterioration in valve function in this group. Gersony and Hayes\(^{21}\) have suggested that aortic valvotomy may increase the risks of infection but since endocarditis is age-related one might expect an increase anyway in the "ageing" postoperative patients. There have been none seen yet in our patients, however, and we doubt if surgery increases the risk of this complication.

Excluding the few problems with aortic regurgitation and the occasional production of mitral regurgitation it appears that there are two courses which patients may take after the first aortic valvotomy. One, slow degenerative calcification, which is relatively simple to manage, and the other with hypoplasia and tunnel obstruction which is difficult and dangerous. The determinant of what happens after aortic valvotomy is the pathological anatomy of the valve at the first operation. Patients at risk of having a difficult reoperation are those with the less common form of congenital aortic valve stenosis described as an atypical variant by Somerville and Ross.\(^{20}\) In our experience this form occurred in 24% of patients; the ring, root, and ascending aorta are small and there is often abnormal subvalvar septal muscle which can form a long obstruction if distal stenosis in the root of valve persists. This type of obstruction resembles the form which occurs in infancy and perhaps represents the milder variety of the same pathology. We think this form is really part of diffuse congenital cardiovascular disease, differing from the more simple and common form of congenital aortic valve stenosis, the pliable dome, which is usually associated with a normal root and post-stenotic aortic dilatation and more often presents later in adolescence.

The type of aortic valve stenosis must be recognised before the first operation, as the operative procedure required may need modification and the ultimate prognosis is different. It is important that those with lumpy valves and hypoplasia of the aorta have good and near complete relief of obstruction; in those below 7 to 8 years we would prefer not to replace the whole root and would therefore recommend gussetting it and leaving some aortic regurgitation to open the outflow. In the few older patients over the age of 10 years who present for the first operation with this form of aortic valve stenosis, aortic root and valve

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**Fig. 6** Peak systolic gradients across the left ventricular outflow tract preoperatively, and postoperatively at rest and after isoprenaline stimulation in nine patients, all of whom had extreme left ventricular hypertrophy on angiography preoperatively. Isoprenaline produced a distinct increase in the gradient in all nine patients.

**Fig. 7** Actuarial survival curves of 49 patients who had open aortic valvotomy: survival at 18 years of follow-up, upper curve reoperation; lower curve no reoperation. Bars show ± standard error at various points.
replacement would now be advised as we feel this procedure provides better long-term relief of the obstruction and is better for the long life of the left ventricle. Unfortunately, these lumpy dysplastic valves are more common in those who present with critical aortic valve stenosis in the first decade. The first operation, which may be regarded as simple, determines the patient’s fate which may be unexpectedly disastrous at reoperation.

Another factor that can contribute to subsequent surgical problems is the unusual disproportionate septal hypertrophy present in some patients, particularly in those with small aortic roots. In our series all these patients had myotomy at the time of valvotomy but we cannot show that myotomy is beneficial as those with and without it are not comparable groups. We think, however, that much more important than performing a myotomy is to leave behind only trivial fixed obstruction. Seventeen patients (35%) have already required reoperation and another one has obvious radiological calcification. Some valve calcification, suggested by the echocardiogram, may be present in a further nine but is not obvious radiologically. The rate of reoperation in patients with calcification is reported in a few series and seems to be related to the length of follow-up. More time must elapse before the complete story is known, but we predict that all will have required reoperation before the end of the second decade. What is needed is something to prevent valve calcification. Restenosis of the aortic valve probably does not occur in the way it does after mitral valvotomy. Obstruction becomes critical from increased rigidity, non-growth of the area, or effects of residual obstruction left at the first operation; this occurred in two patients whose gradients were 80 and 60 respectively after valvotomy and who will require reoperation earlier than the rest. Eight patients who did not need reoperation were followed for more than 10 years and the stenosis had been mildly progressive in four of them. Five of these patients are women who may fare better.

Many reports have claimed good or excellent results for open aortic valvotomy. We accept this if judged by the patient’s early survival, early well-being, and improvement in variables which reflect left ventricular performance. Long-term survival without major problems, however, is now of more concern. At best, open aortic valvotomy can be considered to offer only good palliation, reoperation associated with many problems and some risks being inevitable in the future. The diseased damaged aortic valve remains in the patient and, after valvotomy, the most that can be offered is replacement by a valve with limited life span, known and unknown complications, and inevitable rereplacement. We believe that it is better for a patient to retain, as long as possible, his own aortic valve provided there is no damage occurring to the left ventricular myocardium nor the development of secondary changes which may prejudice the results of reoperation. Thus, aortic valvotomy has an important place in the management of critical aortic valve stenosis, but it is just as important not to do it too early as it is not to do it too late after irreversible myocardial damage.

A knowledge and understanding of the valve pathology before valvotomy may allow more correct prognosis and improve management.

It is also mandatory to establish in the first postoperative year the extent of the residual lesion, keep the patient under regular supervision, and discourage the pursuit of activities which overload the already damaged or “at risk” left ventricular muscle. At the time of the first valvotomy, the future must be seriously considered, knowing that at some time a surgeon must operate again. The goal of aortic valvotomy should be to relieve as completely as possible the obstruction without producing serious aortic regurgitation, and thus improve the life and function of the left ventricle.

References


Addendum

Since submitting this paper for publication in 1980, two more patients have had either valve replacement or valve and root replacement, seven and 15 years after the first valvotomy. One developed progressive calcification and the other with a lumpy valve had a tunnel obstruction.

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