with beta-blockers, angiotensin-converting-enzyme inhibitors, aldosterone antagonists, dual antiplatelet therapy with acetylsalicylic acid and clopidogrel, and statins. An echocardiogram prior to discharge showed a left ventricle ejection fraction of 45%. In this syndrome, ventricular function typically returns to normal levels after several weeks. Our patient lived in another autonomous region and consequently we do not have this datum.

The prognosis of Kounis syndrome is generally good, although complications such as pulmonary edema, arrhythmia and thrombus have been reported during the acute phase. Presentation as cardiogenic shock is extremely rare and only 1 similar case report has been published previously.3

There are no clinical guidelines for treating Kounis syndrome at present. There are too few cases to be able to reach any definitive conclusions on the treatment of this syndrome but, in general, these patients require treatment with corticosteroids, antihistamines, and antithrombotic drugs. Treatment with adrenaline is controversial, as it can worsen ischemia, prolong the QT interval, and induce coronary vasospasm and arrhythmia, but in general it must be administered in the event of severe hypotension or cardiac arrest. Vasodilator agents, including nitrates and calcium channel blockers, must be considered as first-line therapy for previously healthy young patients. The acute coronary syndrome protocol should be followed in patients with the type II variant.4

Kounis syndrome encompasses a group of symptoms that is probably under-diagnosed and should be included in the differential diagnosis of cardiogenic shock.

Most groups opt to correct TGA using the arterial switch before 2 weeks of life. There are anecdotal reports of the procedure being performed beyond 14 days, with the risk of left ventricular failure in the new systemic position.1,2 The two most common causes of this situation are late referral and contraindications for surgery.

Ultrasound criteria for subpulmonary LV exist3 and include paradoxical septal motion, reverse ventricular morphology (spherical right and left semilunar valves), and left free wall thinning, among others. Surgically, a simple measurement of pressures in both ventricles provides a right/left (LV/RV) relationship, with an arterial switch being inadvisable at a ratio < 0.6.

Currently, the 3 main options proposed in this situation are: a) preparatory surgery using pulmonary cerclage and systemic-pulmonary fistula, followed by arterial switch in a second stage;2 b) arterial switch with ECMO support until ventricular remodeling occurs;3,6 c) discharge of the patient and atrial correction (Senning, Mustard) after a few months.6 A priori, the option of using ECMO as support (if necessary) seems the most appropriate.

A German team4 proposed a simple method based on the pressure ratio between the 2 ventricles after a 15–30 min trial period of pulmonary cerclage. After the trial period, the arterial switch is performed if the ratio is adequate (LV/RV > 0.6), or preparation via pulmonary cerclage and fistula is carried out followed by correction in a second stage. The work published by the Great Ormond Street Hospital almost 10 years ago and updated in 20041,5 demonstrated similar survival rates in patients undergoing surgery between 2 weeks and 2 months, and indicated a greater need for inotropic and mechanical ventilation. Recently, another group published results in patients receiving surgery at 6 months,6 and showed higher than usual mortality, despite the use of ECMO. In the

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discuss sections of the article, the authors suggest using an atrial switch (physiological correction) in this subgroup.

None of the children in our series had a prenatal diagnosis. All patients underwent an urgent Rashkind procedure on admission, after diagnosis of TGA (Table 1).

The surgical strategy was based on prioritizing the arterial switch option over preparation (lung cerclage with systemic-pulmonary fistula) if the LV showed no signs of involvment. In cases 1 and 2 we relied on the calculation of intraoperative pressures described by Dabritz et al., as echocardiographic images showed paradoxical movement in the interventricular septum. In case 3, preoperative ultrasound revealed a normal biventricular and septal pattern, so we omitted the pressure calculations.

Cardiopulmonary bypass was carried out with peripheral vasodilatation to decrease the systemic LV afterload. Epicardial ultrasound-guided cardiopulmonary bypass output was therefore gradual, and was used for circulatory assistance, with vasodilator support (levosimendan, solinitrin) and adrenaline, and systolic pressures not exceeding 50 mmHg. The chest was left open prophylactically. Data from the immediate postoperative period barely differed from other common arterial switches (Table 2). LV contractility was considered normalized within 48 h. Peritoneal dialysis was required in cases 2 and 3 (the latter also required preoperative dialysis). The 3 children were extubated on the fourth day after surgery and remained in intensive care for between 6 and 8 days.

In conclusion, because of late referrals or transient formal contraindications, performing an arterial switch beyond 2 weeks may have to be considered. Intraoperative ultrasound and pressure criteria can help to decide between initial corrective surgery or preparation and correction in 2 stages. An aggressive strategy of perioperative and postoperative peripheral vasodilatation probably facilitates left ventricular remodeling. In our brief experience, recovery was rapid and complete, without the need for postoperative ECMO.

Table 1
Preoperative Data

<table>
<thead>
<tr>
<th>Age on admission, days</th>
<th>Emergency Rashkind</th>
<th>Age at surgery, days</th>
<th>Weight, g</th>
<th>Diagnosis</th>
<th>Complications</th>
<th>Aristotle score</th>
</tr>
</thead>
<tbody>
<tr>
<td>18</td>
<td>Yes</td>
<td>19</td>
<td>3100</td>
<td>Simple TGA, situs inversus</td>
<td>Shock</td>
<td>25</td>
</tr>
<tr>
<td>1</td>
<td>Yes</td>
<td>27</td>
<td>3280</td>
<td>Simple TGA</td>
<td>Shock, sepsis, acute renal failure</td>
<td>22</td>
</tr>
<tr>
<td>6</td>
<td>Yes</td>
<td>28</td>
<td>3200</td>
<td>Simple TGA</td>
<td>Shock, sepsis, acute renal failure</td>
<td>24</td>
</tr>
</tbody>
</table>

TGA, transposition of the great arteries.

Table 2
Pre- and Postoperative Data

<table>
<thead>
<tr>
<th>LV/RV, %</th>
<th>Extracorporeal circulation, min</th>
<th>Aortic clamping min</th>
<th>ICU lactate</th>
<th>Open chest, days</th>
<th>Intubation, h</th>
<th>ICU, days</th>
</tr>
</thead>
<tbody>
<tr>
<td>45/55 (0.82)</td>
<td>149</td>
<td>68</td>
<td>3.9</td>
<td>1</td>
<td>96</td>
<td>6</td>
</tr>
<tr>
<td>40/50 (0.80)</td>
<td>180</td>
<td>70</td>
<td>8.1</td>
<td>2</td>
<td>96</td>
<td>7</td>
</tr>
<tr>
<td>40/50 (0.80)</td>
<td>160</td>
<td>59</td>
<td>3.8</td>
<td>1</td>
<td>96</td>
<td>8</td>
</tr>
</tbody>
</table>

ICU, intensive care unit; LV, left ventricle; RV, right ventricle.

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