Unknown Complication of Arterial Switch Operation
Resistant Hypertension Induced by a Strong Aortic Arch Angulation

Magalie Ladouceur, MD; Pierre Boutouyrie, MD, PhD; Younes Boudjemline, MD, PhD; Hakim Khettab, PhD; Alban Redheuil, MD, PhD; Antoine Legendre, MD; Sarah Cohen, MD; Laurence Iserin, MD; Damien Bonnet, MD, PhD; Elie Mousseaux, MD, PhD

A 17-year-old man with complete transposition of the great arteries who had undergone the arterial switch procedure was referred to our tertiary center to investigate systemic hypertension. High blood pressure (BP) was first documented at age 13 years and was controlled until age 16 years by enalapril (20 mg/d) and atenolol (100 mg/d). BP became uncontrolled despite the addition of 2 other antihypertensive drugs (amlodipine 10 mg/d and furosemide 20 mg/d).

At admission, hypertension was still observed despite treatment. Left arm BP was 141/64 mm Hg, right arm BP was 149/63 mm Hg, left inferior limb BP was 153/58 mm Hg, and right inferior limb BP was 155/60 mm Hg. Twenty-four-hour ambulatory BP monitoring confirmed hypertension, with mean daytime BP at 159/78 mm Hg. Initial laboratory investigations, including measures of serum potassium and creatinine, quantitative proteinuria, renin, aldosterone, corticosteroids, and catecholamines in plasma and urine, were normal. Renal and adrenal ultrasound scans were also normal. Echocardiography showed moderate left ventricular (LV) hypertrophy. The aortic root was dilated, and aortic regurgitation was graded as mild (Table). An instantaneous gradient of 36 mm Hg was measured with a continuous-wave sample through the aortic isthmus. Cardiovascular magnetic resonance angiography performed with a 1.5-Telsa magnet showed a common origin of the left and right carotid arteries from the brachiocephalic artery, as well as a visible narrowing of the descending aorta (minimum diameter 14 mm) insufficient to define coarctation. A major finding was that the aortic arch had a Gothic shape with a very marked 73° angulation between the ascending and descending aorta (Figure 1; Movies I and II in the online-only Data Supplement). LV ejection fraction was normal in cardiovascular magnetic resonance angiography, but concentric LV hypertrophy was confirmed (Table). To better understand the cause of hypertension, we carefully measured peripheral BP in laboratory conditions and central BP using applanation tonometry (SphygmoCor, AtCor Medical, Sydney, Australia). BP was measured simultaneously in both arms by use of similar automated brachial artery sphygmomanometers (Colin 8800 P, Colin Corporation Ayashi, Komaki, Japan), with the devices alternated between each set of measurements. A significant and consistent difference of 10 to 12 mm Hg between the right and left arm was observed, with higher BP measured in the right arm. The applanation tonometry was performed according to guidelines.1 An early reflection wave was observed both on the right and left carotid artery waveform, 60 ms after systole onset, followed by the usual reflection wave that occurs in late systole (Figure 2).

Applanation tonometry was performed immediately afterward on the femoral artery. Pulse-wave velocity was paradoxically low at 4 m/s, which corresponds to the first percentile of the expected value according to age.1 This value of pulse-wave velocity enabled us to calculate the location of the reflection site of the early wave reflection, estimated to be 24 cm from the right carotid artery. When reported on aortic anatomy in cardiovascular magnetic resonance angiography, this could correspond to the aortic angulation site.

Aortic pressures were measured invasively. These pressures were 108/45/70 mm Hg in the descending aorta, 116/44/74 mm Hg in the aortic arch just after the aortic angulation, and 135/43/71 mm Hg in the ascending aorta just before the aortic angulation. Peak-to-peak gradient was estimated at 19 mm Hg, which could favor a functional aortic stenosis, but conventional angiography confirmed the cardiovascular magnetic resonance findings of the absence of coarctation along this folded aortic arch (Figure 1). The early reflection wave added to the incident systolic wave was observed on the pressure curve in the ascending aorta, which led to augmentation of the systolic pressure from 110 to 135 mm Hg (Figure 3), whereas in the descending aorta, pressure-curve morphology was normal.

We illustrate here a case of resistant hypertension, the pathogenesis of which could be related to a highly angulated aortic arch. Although excessive wave reflection could easily explain LV hypertrophy in our patient, because the LV is directly exposed to increased BP, the relation with systemic...
Hypertension is not trivial. The early reflection of the systolic wave in the ascending aorta, the reflection site of which could match the strong angulation of the Gothic-shaped aortic arch, increased the systolic pressure by addition of the incident and reflected waves (Figures 2 and 3). Because the angulation was very localized between the brachiocephalic trunk (the common origin of the left and right common carotid arteries) and the left subclavian artery, the right brachial and carotid pressures were higher than the left brachial pressure. In contrast, BP in the descending aorta was in the normal range because the early reflection wave did not affect this vessel (Figures 2 and 3).

Although the local peak-to-peak invasive pressure gradient across the angulation was measured at 19 mm Hg, the true difference was larger and estimated to be 27 mm Hg between the ascending and descending aorta.

The relationship between the Gothic-shaped aortic arch and early pulse-wave reflection, hypertension, and LV hypertrophy has been demonstrated after aortic coarctation repair without significant stenosis. In patients who have undergone the arterial switch operation, aortic arch angulation has been found to be more acute and associated with early pulse-wave reflection in the pediatric population. This geometry results from root mobilization during the arterial switch operation and in the present case was accentuated by pressure of the pulmonary artery bifurcation on the ascending aorta, which led to a kink of the arch (Figure 1). This complication of the arterial switch operation can be induced when the pulmonary arteries are not sufficiently dissected, and therefore are flexible, during the Lecompte maneuver. Patients who have undergone the arterial switch operation should be screened for hypertension, especially during adolescence. Given the refractory nature of hypertension in our patient, and if aortic arch geometry is causal, adaptation of the surgical procedure aimed at preserving the Romanesque architecture is warranted.

### Disclosures

None.

### References


### Table. Echocardiographic and CMR Measures

<table>
<thead>
<tr>
<th>Measures</th>
<th>Echocardiography</th>
<th>CMR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diastolic IVS thickness, mm</td>
<td>11</td>
<td>12</td>
</tr>
<tr>
<td>LV posterior wall thickness, mm</td>
<td>11</td>
<td>12</td>
</tr>
<tr>
<td>Diastolic LV diameter, mm</td>
<td>54</td>
<td>56</td>
</tr>
<tr>
<td>Indexed LV mass, mg/m²</td>
<td>130</td>
<td>95.5*</td>
</tr>
<tr>
<td>LVEF, %</td>
<td>72</td>
<td>71</td>
</tr>
<tr>
<td>Hypertensive diastolic pressure, mm</td>
<td>37 (or 19 mm/m²)</td>
<td>37.5×37.5</td>
</tr>
<tr>
<td>Aortic diameter, mm</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sinus of Valsalva</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ascending aorta</td>
<td></td>
<td>21</td>
</tr>
<tr>
<td>Aortic arch</td>
<td></td>
<td>14</td>
</tr>
<tr>
<td>Aortic isthmus</td>
<td></td>
<td>15</td>
</tr>
<tr>
<td>Aortic regurgitation grade</td>
<td>Mild</td>
<td>Mild (RF=17%)</td>
</tr>
</tbody>
</table>

CMR indicates cardiovascular magnetic resonance; EF, ejection fraction; IVS, interventricular septum; LV, left ventricular; and RF, regurgitation fraction.

*LV mass is higher than the mean value +2 SD estimated in healthy male volunteer in our center.
Figure 1. Magnetic resonance angiography of the thoracic aorta (A) and conventional catheterization (B) showed a strong angulation of the aortic arch (angle =73°; A), just after the common origin of the innominate artery and left common carotid artery.

Figure 2. Applanation tonometry on the carotid arteries and the radial arteries. An early reflection wave (*) was observed on the waveform of the right and left carotid artery and on the waveform of the right radial artery. Arrows indicate usual diastolic reflection wave. The early reflection wave occurred 60 ms after systole onset in the right carotid artery and 144 ms after systole onset in the right radial artery. With pulse-wave velocity measured at 4 m/s, the reflection site of the early wave reflection was calculated to be located 24 cm from the right carotid artery and 57 cm from the right radial artery, which could correspond with the aortic angulation site. Left radial waveform morphology was normal.

Figure 3. Pressure curve in the ascending aorta (Asc Ao; left) and descending aorta (Desc Ao; right) during aortic catheterization. The early reflection wave (*) added to the incident systolic wave (red arrows) was observed on the pressure curve in the ascending aorta, which led to augmentation of systolic pressure from 110 to 135 mm Hg, whereas in the descending aorta, peak systolic pressure was much lower (108/45/70 mm Hg), with the reflection wave occurring late in systole (black arrow).
Unknown Complication of Arterial Switch Operation: Resistant Hypertension Induced by a Strong Aortic Arch Angulation
Magalie Ladouceur, Pierre Boutouyrie, Younes Boudjemline, Hakim Khettab, Alban Redheuil, Antoine Legendre, Sarah Cohen, Laurence Iserin, Damien Bonnet and Elie Mousseaux

_Circulation_. 2013;128:e466-e468
doi: 10.1161/CIRCULATIONAHA.113.002097

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2013 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/128/25/e466

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2014/03/20/128.25.e466.DC1

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/
**Movie Legend**

SSFP cine images of the aortic arch including both ascending and descending aorta in a sagittal oblique view (movie 1) and in coronal oblique view along the LV outflow tract and the ascending aorta (movie 2). Severe angulation between the ascending and descending segment of the thoracic aorta is well shown as well the high systolic pulsatility of the proximal aorta. Strong back flow occurring during diastolic recoil of the aortic wall can also be well detected in the ascending aorta while the aortic valve regurgitation was mild. The recommended application for viewing the movie 1 and 2 is Windows Media Player.