Cardiological and general health status in preschool- and school-age children after neonatal arterial switch operation

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Abstract

Objective: Cardiological and general health status 3–9 years after neonatal arterial switch operation for transposition of the great arteries should be evaluated by non-invasive methods. Methods: A total of 77 unselected children with intact ventricular septum (75.3%) or ventricular septal defect (24.7%) without or with aortic isthmic stenosis (5.2%) were prospectively examined 3.2–9.4 years (5.4±1.6) after neonatal switch. Clinical pediatric and cardiological examination, standard and 24 h Holter electrocardiogram, M-mode, 2D-, Doppler and colour Doppler echocardiography were performed. Outcome data were compared to published normals. Results: Reoperation rate was 2.6%, 96.1% were without limitation of physical activity and 98.7% without medication. Compared to normals, growth was adequate, weight and head circumference were slightly reduced. After median sternotomy, 23.4% had abnormal thoracic configuration (16.9% asymmetry, 6.5% funnel chest). ECG and Holter: 93.5% were in sinus, 6.5% in ectopic atrial or junctional rhythm. Incidence of complete right bundle branch block was 15.8% in patients with ventricular septal defect and 5.2% in those without. Ischemic ST-T changes during exercise due to coronary artery occlusion and evidence of old myocardial infarction were found in 1 patient (1.3%) each.occasional atrial ectopy was found in 27.4%, ventricular ectopy in 15.3%: occasional in 12.5% and frequent (>30/h) in 2.8% presenting bigemini, couplets and short runs of ventricular tachycardia at rest and during exercise. Echocardiography: Left ventricular function was normal in all. Endystolic diameter of neoaortic valve annulus was beyond 90% confidence interval for controls in 79.2%, neoaortic root diameter in 100%. Mild aortic insufficiency was seen in 10.4%. No correlation was found between aortic insufficiency and aortic dilatation. Neoaortic stenosis was not seen, mild residual coarctation after end-to-end-anastomosis was found in 2.6%, native coarctation corrected later on in 1.3%. Supravalvular pulmonary stenosis was seen in 29.9% (19.5% trivial, 7.8% mild, 2.6% moderate), mild subvalvular pulmonary stenosis in 1.3%, pulmonary insufficiency in 2.6%. Conclusion: The study confirms good midterm results after neonatal arterial switch operation for transposition with or without ventricular septal defect. Long-term observation is necessary to assess rhythm, coronary artery and myocardial function as well as development of neo-aorta and pulmonary artery system. © 1997 Elsevier Science B.V.

Keywords: Complete transposition; Arterial switch operation; Electrocardiogram; Holter monitoring; Echocardiography

1. Introduction

The neonatal arterial switch operation is currently the surgical procedure of choice for correction of transposition of the great arteries with or without ventricular septal defect [2,14,20,24,25,28,29,34–36]. The advan-
tages of anatomic correction over the physiological repair of Mustard and Senning with respect to arrhythmias and function of the systemic ventricle have been supported again in several recent follow-up studies [3,21,26,32].

The purpose of the present study was to review a 6-year experience in neonates who had undergone an arterial switch operation for treatment of transposition of the great arteries with intact ventricular septum or ventricular septal defect, as well as accompanying coarctation of the aorta in our institution. Report is given on a prospective follow-up study performed 3–9 years after surgery in order to evaluate by non-invasive methods cardiological findings and general health status of the children, whereas a recent study concerned neurological and developmental outcome of the same patients [12].

In addition, information should be obtained as to which extent certain peri- and postoperative factors correlate to later outcome parameters.

2. Materials and methods

2.1. Demographic data

Between March 1986 and February 1992, 96 consecutive newborns with transposition of the great arteries (TGA) underwent arterial switch operation (ASO) in our institution. There were 89 long-term survivors.

The study group consisted of 77 unselected children (86.5% of the survivors), whose parents consented to bring their child back to this prospective follow-up examination. Of the children 55 were male (71%) and 22 were female (29%). Birth weight ranged from 2.1 to 5.4 kg (3.49 ± 0.5 kg, mean ± S.D.).

A total of 55 (71.4%) had a simple TGA, in addition 15 (19.5%) also had a non important ventricular septal defect (VSD) not closed during ASO, 3 (3.9%) a VSD closed during ASO, and 4 (5.2%) a coarctation of the aorta corrected later on. In summary, 75.3% of the patients had an intact ventricular septum and 24.7% a VSD. Two patients (2.6%) had a non-stenotic dysplastic (bicuspid) pulmonary valve.

Coronary artery status according to [7] was usual (type AI) in 62 (80.5%) of the studied patients. In 4 cases (5.2%), the circumflex coronary artery arose from the right coronary artery (type ABI), in 5 cases (6.5%) right coronary artery and circumflex coronary artery were inverted (type AIII); in four (5.2%) there was a single ostium for the left and right coronary arteries from the right posterior facing sinus (type BI), and in two (2.6%) there was a single ostium for the left and right coronary arteries from the left anterior facing sinus (type AII).

Prostaglandin E1-infusion (0.025–0.1 μg/kg/min) was administered until the operation in 97.4% to keep the arterial duct open. Preoperative heart catheterization was performed in 93.5%, and balloon-atrio-septostomy in 84.4% of the patients.

2.2. Surgical management

In the study group, the age at ASO ranged from 2 to 39 days (7.0 ± 4.6), 2 patients were older than 12 days. Surgery was performed under deep hypothermic circulatory arrest (DHCA) with an oesophageal temperature of 14–17°C (15. ± 1.0°C) and combined low flow cardiopulmonary bypass (CPB). Total CPB time ranged from 41 to 116 min (63.7 ± 15.2), DHCA time ranged from 51 to 70 min (60.1 ± 4.5). The mean aortic cross clamp time was 66.1 ± 8.5 min.

Standardized surgical techniques for the ASO, with or without ventricular septal defect closure, were based on the Lecompte-modification [15] to establish right ventricle to pulmonary artery continuity. The proximal neo-pulmonary trunk was reconstructed with two autologous pericardial patches filling the coronary artery excision sites. The atrial septal defect was closed by direct suture in all but 1 patient, the patent ductus arteriosus was ligated and divided in all patients.

One patient with an usual coronary artery pattern (type AI) needed immediately after ASO a mammaria interna bypass operation to the right coronary artery because of myocardial infarction caused by intraoperative damage to that artery [8]. Another patient needed reconstruction of the tricuspid valve after valvular damage during the Rashkind procedure.

2.3. Follow up study protocol

The evaluation of the 77 unselected patients participating in the prospective study was performed at an age of 3.2–9.4 years (5.4 ± 1.6).

The protocol included an interview with the child and its accompanying persons as well as clinical pediatric and cardiological examination performed by one pediatric cardiologist.

A 12-lead standard electrocardiogram (ECG) at rest was performed. Standard age corrected criteria for chamber hypertrophy and conduction intervals [4] were applied. Rhythm and repolarisation abnormalities were analyzed applying generally accepted criteria [6].

Two channel 24 h Holter monitoring was performed, minimal and maximal heart rates and rhythm were evaluated by means of computer-assisted analysis complemented by manual examination of printouts [30,31].

M-mode and two-dimensional echocardiography, Doppler and colour Doppler echocardiography were performed with an ATL-HDI 3000 CV or ATL-Ultrasound mark 800 unit. Standard long axis views were used to
visualize the neo-aorta and the ventricles. Pulsed and continuous wave Doppler were applied to assess valvular competence and to identify obstructions of the great vessels or their branches. Colour Doppler was used to examine aortic insufficiency or intracardiac shunts.

Left heart dimensions and shortening fraction were compared to those of normal children [9].

Aortic diameters were compared to those of normal children [10]. Still frames were taken from the parasternal long axis view at the end of systole. The internal diameter of the valvular annulus was measured at the hinge point of the valve leaflets, the internal diameter of the aortic root at its maximal width.

Doppler peak instantaneous gradients were calculated using the simplified Bernoulli equation. Supravalvular pulmonary stenosis was classified by Doppler gradients as absent (up to 16 mmHg), trivial (17–24 mmHg), mild (25–39 mmHg), moderate (40–59 mmHg) and severe (more than 59 mmHg) [27].

Insufficiency of the neo-aortic valve (AI) was quantified by colour Doppler as absent (0 mm), mild (1–3 mm diastolic backflow), moderate (4–6 mm diastolic backflow) or severe (more than 6 mm diastolic backflow) [11,13].

2.4. Statistical analysis

The outcome data included both categorical and continuous variables. They were statistically related to categorical and continuous peri- and postoperative variables. The Fisher exact test was used to analyze categorical variables. Correlation analyses with Pearson correlation coefficients were performed between continuous variables.

P-values < 0.05 were considered significant.

3. Results

3.1. Reoperation rate and health status

Of the 77 patients, 6 (7.8%) had undergone a second intervention during follow-up. In 4 of these, an aortic coarctation had been corrected at age 1 month to 4 years. One patient had undergone resection and patch plastic reconstruction of a severe right ventricular outflow tract (RVOT) obstruction at the age of 3 years. Another asymptomatic patient (coronary pattern type BI) had needed a mammaria interna bypass operation to the proximal anterior descending coronary artery at the age of 5 years after complete occlusion of the main left coronary artery with retrograde perfusion of its branches via collaterals from the right coronary system.

Of the 77 patients, 74 (96.1%) had no limitation of physical activity (class I following the New York Heart Association-NYHA); 2 patients (2.6%) showed mild limitation (class II): the patient after neonatal bypass operation following myocardial infarction [8] suffering from nodal rhythm and severe ventricular ectopic activity, and another with moderate pulmonary valve insufficiency. One patient could not be staged because of neurological impairment.

A total of 76 patients (98.7%) were without cardiac medication, and digoxin therapy was terminated at the time of the follow-up examination in one 4-year-old asymptomatic girl who had Wolff–Parkinson–White syndrome.

Growth of the 77 patients was normal compared to the normal population [1] (Table 1). In contrast, weight development was slightly reduced in comparison to normal children. Statistical analysis revealed a significant correlation between low weight (<10. percentile) at birth and at the time of follow-up (P = 0.02) suggesting that reduced weight is not related to ASO. Fronto-occipital head circumference was reduced compared to the normal: microcephaly (<3. percentile in 4 patients) was due to hypoxic cerebral damage in two, to velo-facio-cardiac syndrome in one and to embryo-fetal alcohol syndrome in the remaining patient. Systolic and diastolic arterial blood pressures at rest were normal in 98.7% of the children. One patient had slightly elevated systolic and diastolic arm pressures due to his native coarctation of the aorta.

None of the children had signs of cardiac insufficiency.

Systolic heart murmurs were noted in all patients, usually at the base of the heart. 18.2% had murmurs grade I, 66.2% grade II, 11.7% grade III and 3.9% grade IV. Grade III and grade IV murmurs were associated with hemodynamically non important VSD (n = 5)
Table 2
Standard ECG-findings after neonatal ASO

<table>
<thead>
<tr>
<th></th>
<th>Follow up 5.4 ± 1.6 years (n = 77)</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Rhythm</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sinus</td>
<td>72</td>
<td>93.5</td>
</tr>
<tr>
<td>Ectopic atrial</td>
<td>2</td>
<td>2.6</td>
</tr>
<tr>
<td>Junctional</td>
<td>3</td>
<td>3.9</td>
</tr>
<tr>
<td><strong>Intraventricular conduction</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal</td>
<td>33</td>
<td>42.9</td>
</tr>
<tr>
<td>Incomplete RBBB</td>
<td>38</td>
<td>49.4</td>
</tr>
<tr>
<td>Complete RBBB</td>
<td>6</td>
<td>7.8</td>
</tr>
<tr>
<td></td>
<td>3 (IVS)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3 (VSD)</td>
<td></td>
</tr>
<tr>
<td><strong>Ventricular hypertrophy</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>66</td>
<td>85.8</td>
</tr>
<tr>
<td>LVH</td>
<td>2</td>
<td>2.6</td>
</tr>
<tr>
<td>RVH</td>
<td>9</td>
<td>11.7</td>
</tr>
<tr>
<td><strong>Atrial hypertrophy</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>77</td>
<td>100.0</td>
</tr>
<tr>
<td><strong>Ventricular ectopic activity</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>74</td>
<td>96.1</td>
</tr>
<tr>
<td>VPB single</td>
<td>1</td>
<td>1.3</td>
</tr>
<tr>
<td>Bigemini</td>
<td>1</td>
<td>1.3</td>
</tr>
<tr>
<td>VT (short runs up to 5 consecutive beats)</td>
<td>1</td>
<td>1.3</td>
</tr>
<tr>
<td>Evidence of myocardial infarction</td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>76</td>
<td>98.7</td>
</tr>
<tr>
<td>Present</td>
<td>1</td>
<td>1.3</td>
</tr>
<tr>
<td><strong>ST-T-Wave Changes</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>76</td>
<td>98.7</td>
</tr>
<tr>
<td>Present</td>
<td>1</td>
<td>1.3</td>
</tr>
</tbody>
</table>

* 5.2% of the patients with intact ventricular septum (IVS) (n = 58);
* 15.8% of the patients with ventricular septal defect (VSD) (n = 19);

RBBB, right bundle branch block; LVH, left ventricular hypertrophy; RVH, right ventricular hypertrophy; VPB, ventricular premature beats; VT, ventricular tachycardia.

or supravalvular pulmonary stenosis (n = 7) of mild to moderate degree.

Diastolic heart murmurs were noted in 2 patients (2.6%) who had pulmonary insufficiency; no patient with neo-aortic valve regurgitation as assessed by echocardiography had a diastolic heart murmur.

Asymmetry of the right and left hemithoraces with visible prominence of the left ventral ribs was found in 12 (16.9%) of the patients. Of these 4 patients received orthopedic treatment and physiotherapy. There were 5 patients (6.5%), all without hereditary taint, who showed a mild funnel chest.

In total, 18 (23.4%) of the patients showed a striking thoracic configuration after neonatal median sternotomy, whereas the scars were smooth and sufficiently adapted in all but 3 patients.

3.2. Surface electrocardiography (ECG) at rest (Table 2)

3.2.1. Rhythm

Of the study patients, 72 (93.5%) were in sinus rhythm, 2.6% had an ectopic atrial rhythm, and 3.9% a junctional rhythm. One patient without history of tachycardia showed Wolff–Parkinson–White syndrome. There was no patient with first or second degree or complete heart block. No patient needed a pacemaker. Statistical analysis showed a significant correlation between arrhythmias in the ECG (atrial or junctional rhythm) obtained at hospital discharge (27 ± 21 days after ASO) and at the time of the follow up study (P = 0.008).

There were 3 patients who had monomorphous ventricular ectopic beats in the standard ECG: one of them (coronary pattern ABI) showed isolated ventricular premature beats (VPB), another (type AI) had ventricular bigemini and the third (type BI) couplets and short runs up to 4 beats of ventricular tachycardia (VT). These latter 2 patients also had important ventricular ectopy on Holter monitoring and are described in more detail under this heading. Atrial premature beats (APB) were not noticed.

No statistical correlation was found between usual or unusual coronary artery patterns and ventricular ectopic activity at rest.

3.2.2. Intraventricular conduction abnormalities

So called incomplete right bundle branch block (RBBB) in form of Rs'-patterns with normal QRS-duration was seen in 38 patients (49.4%), complete RBBB in 6 patients (7.8%). Of the latter, 3 patients had a VSD which was closed during ASO in two. The incidence of complete RBBB was 5.2% in TGA without, and 15.8% in TGA with ventricular septal defect.
3.2.3. **Ventricular hypertrophy**

One patient with a concomitant native coarctation of the aorta met ECG criteria for left ventricular hypertrophy.

A total of 9 patients (11.7%) had signs of right ventricular hypertrophy; 6 of these had mild to moderate supravalvular pulmonary stenosis, one moderate right ventricular outflow tract obstruction, and one pulmonary valve insufficiency.

3.2.4. **ST-T wave changes, myocardial infarction**

The patient with VT had signs of an old posterior wall infarction which had occurred during ASO and had necessitated an arteria mammaria interna bypass to the right coronary artery at the same operative session. None of the other patients (the 6 with complete RBBB excluded) had evidence of myocardial infarction or ischemia at rest.

3.3. **24-h ambulatory ECG monitoring**

Follow-up Holter monitoring was available in 72 (93.5%) of the study patients at an age of 3.2 to 9.4 years (5.4 ± 1.6) (Table 3).

3.3.1. **Rhythm**

Sinus rhythm was present in 68 cases (94.5%) and short periods of junctional rhythm alternating with sinus rhythm in 4 (5.5%). In the patient with Wolff–Parkinson–White syndrome this was present throughout the 24 h.

3.3.2. **Rate**

The minimal heart rate averaged 53 ± 8 (range 36–80) beats/min, the maximal heart rate 167 ± 22 (range 89–210) beats/min. The mean heart rate at sleep (measured during at least 5 sleeping h) was 61 ± 9 (range 42–90) beats/min. One patient had short sinus pauses (2.1–2.4 s) during sinus bradycardia at sleep.

3.3.3. **Atrial ectopy**

A total of 19 patients (26.0%) had occasional and 1 patient (1.4%) frequent (more than 15/h) atrial ectopic beats. Episodes of supraventricular tachycardia were not noticed.

3.3.4. **Ventricular ectopy**

Ventricular ectopy was present in 11 patients (15.3%). Occasional ventricular ectopic beats were found in 9 (12.5%) children while 2 (2.8%) had frequent (more than 30/h) ventricular ectopic beats. In one of these latter patients with prevailing junctional rhythm bigemini, couplets as well as short runs of ventricular tachycardia (3–4 beats) were present at rest and during exercise. This patient, 6.8 years of age, with a coronary pattern type AI had, as already mentioned, sustained a myocardial infarction immediately after ASO and therefore received a mammaria interna bypass to the right coronary artery [8]. Antiarrhythmic therapy was not successful. The other patient, 4.2 years of age, with a coronary pattern type BI, showed frequent ventricular ectopic activity, bigemini and couplets as well as short runs of ventricular tachycardia (4–5 beats) at rest and during exercise. Coronary artery stenosis was excluded by coronarography. Antiarrhythmic therapy with mexiletin was successful in reducing ectopic activity.

3.3.5. **Ischemic ST-T wave changes**

There was one patient (follow-up 4.5 years, coronary pattern type BI) with ischemic changes of the ST-T waves during exercise (heart rate > 170 beats/ min). Coronarography revealed occlusion of the left anterior descending coronary artery with retrograde perfusion of the vessel from the right coronary artery system. This patient underwent mammaria interna bypass to the left coronary artery at the age of 5 years. No statistical correlation was found between the coronary artery pattern and dysrhythmias on Holter monitoring.

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Table 3

<table>
<thead>
<tr>
<th>Holter monitor findings after neonatal ASO</th>
<th>Follow-up 5.2 ± 1.5 years (n = 72)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rhythm</td>
<td>Sinus</td>
</tr>
<tr>
<td></td>
<td>Junctional</td>
</tr>
<tr>
<td>Atrial ectopic activity</td>
<td>No</td>
</tr>
<tr>
<td></td>
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<td></td>
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<td>Ventricular ectopic activity</td>
<td>No</td>
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<td></td>
<td>Occasional (&lt;30 beats/h)</td>
</tr>
<tr>
<td></td>
<td>Frequent (&gt;30 beats/h), bigemini, couplets, ventricular tachycardia (up to 5 consecutive beats)</td>
</tr>
</tbody>
</table>
3.4. Echocardiography (Table 4)

3.4.1. Left ventricle

M-mode recordings showed a normal left ventricular enddiastolic diameter (LVEDD) in 71 patients (92.2%). In 1 patient (1.3%) this diameter was below the 3rd percentile, in 5 (6.5%) above the 97. percentile.

The shortening fraction (SF) ranged from 29 to 52% (mean 39 ± 5%). In no patient was it below the 3rd percentile (< 28%), 5 patients (6.5%) had values above the 97. percentile (> 47%).

3.4.2. Dimensions of the neo-aorta

The endsystolic neoaortic valve annulus diameter ranged from 16 to 31 mm (22 ± 4). The estimated mean diameter was calculated 41.2% higher than that for age-matched normal children [10], representing an average augmentation in diameter of 6 mm (Fig. 1, regression lines). 79.2% of the patients had diameters beyond the 90% confidence interval for control subjects, 20.8% were within the limits.

The endsystolic neoaortic root diameter ranged from 24 to 44 mm (33 ± 5). The estimated mean diameter was calculated 92.4% higher than that for age-matched normal children [10], representing an average augmentation in diameter of 16 mm (Fig. 2, regression lines). All patients had diameters beyond the 90% confidence interval for control subjects.

3.4.3. Neo-aortic valve insufficiency (AI)

Neo-aortic valve regurgitation was detected in 8 patients (10.4%), whereas no AI was found in 69 patients (89.6%). In each case, regurgitation was of mild degree, only present in the immediate subaortic region.

No correlation was found between the occurrence of AI and the degree of neo-aortic valve annulus or neo-aortic root dilatation.

3.4.4. Neoaortic stenosis

Peak Doppler gradients across the aortic valve (5.7 ± 2.3 mmHg, patients not sedated) were normal in all patients and showed no signs of stenosis, independent of a bi- or tricuspid valve.
with gradients between 17 and 24 mmHg; 6 (7.8%) mild SPS with gradients between 25 and 39 mmHg, and 2 asymptomatic patients showed moderate SPS with gradients of 44 and 56 mmHg, respectively (patients not sedated). Heart catheterization was performed in both (follow-up 6.9 and 7.5 years) and showed kinking at the site of the anastomosis with peak to peak gradients of 20 and 40 mmHg. Reoperation was not deemed indicated at present in both cases. In addition, one patient (follow-up 9.4 years) had mild residual subvalvular stenosis with a gradient of 30 mmHg after resection and patch plastic reconstruction of a severe right ventricular outflow tract obstruction at the age of 3 years.

3.4.7. Pulmonary insufficiency

Pulmonary valve insufficiency (PI) was present in 2 patients (2.6%). It was mild in one and of moderate degree in the other; surgical reintervention was not considered to be indicated at the present time.

4. Discussion

A distinguishing feature of the present study is the fact that its results are based on a very careful prospective examination carried out by one of us (H.H. Hövels-Gürich) during a period of 11 months. Reintervention after ASO was rare in our study group, and compared to other series [2,16,24,28,33,34], reoperation rate for pulmonary stenosis (1.3%) was the lowest one in our patients.

Asymmetry of the right and left hemithoraces after neonatal median sternotomy in 16.9% of our patients indicating orthopedic treatment and physiotherapy aiming to prevent scoliosis is an important observation from the pediatric point of view, and careful further clinical investigations of children after this type of surgical access at neonatal age are required.

General pediatric health status was very satisfying and not different compared to normal children. While intelligence was normal, the incidence of neurological and motor dysfunctions was higher than in an age-matched control population [12].

The advantages of anatomic over atrial repair of TGA with respect to the preservation of sinus node function and a low incidence of significant atrial or ventricular rhythm disturbances are well documented by our and by several previous studies [16,18,20,21,23,26,28,32,33].

Ventricular ectopy was rare in our study group compared to other studies [21,26]. Severe ventricular ectopy such as bigeminy, couplets or runs of ventricular tachycardia may be an indicator for ventricular dysfunction or coronary insufficiency. This fact emphasizes the importance of repeated ECG examination and exercise testing and careful search for the etiology of ventricular ectopy including coronary angiography.
Ischemic ST-T-wave changes, occurring at exercise in asymptomatic patients, must be considered as an additional indicator of insufficient coronary blood supply requiring coronary angiography. As severe ventricular ectopy and ischemic changes at rest or during exercise are suspected to occur secondary to reduced coronary flow reserve, a relation to the coronary artery morphology may be assumed and was reported in a former study [35]. In our series, we found no significant correlation between severe ventricular ectopy and coronary artery status.

Left heart morphologic and functional sequelae are rare following anatomic correction of TGA. Follow-up periods up to 10 years reveal no evidence of time-related deterioration of left ventricular function [3,23,28,29,32,33].

Our data demonstrate significantly increased diameters of the neo-aortic valve annulus and root compared to normal children, confirming the results described in 2 recent studies [11,29]. In contrast to [11], we could not find a correlation between the degree of aortic root dilatation and the occurrence of neo-aortic valve regurgitation in our patients. We confirm, however, the observation that no correlation exists between the degree of neo-aortic valve annulus dilatation and aortic insufficiency.

The incidence of neo-aortic valve regurgitation (10.4%) in our patients as assessed by colour Doppler was quite low and its degree was always mild. The fact that mild pulmonary valve regurgitation is detected in a high percentage (28–88%) of normal patients of various ages by colour Doppler [36] indicates that this might be a normal finding not indicative of pathology, as colour Doppler is very sensitive in detecting mild valvular regurgitations. Further follow-up studies are necessary to determine whether mild neo-aortic valve regurgitation will eventually develop clinical or hemodynamic significance.

In our study group as in other reports [2,5,16,17,22,23,28,29,33–35], supravalvular pulmonary stenosis is the most common postoperative complication after ASO. Its incidence ranges from 8 to 48%, the definition of severity changing from author to author and depending on the method applied (catheter gradients versus Doppler gradients). Mild systolic gradients between right ventricle and pulmonary artery are common, but the development of a severe supravalvular pulmonary stenosis (Doppler gradients higher than 59 mmHg) is rare and ranges from 1 to 10% in the literature.

Various mechanisms have been implicated in the development of supravalvular pulmonary stenosis. Most of the increase of the gradient between right ventricle and the pulmonary artery system occurs during the first year after repair when somatic growth is especially high. Circumferential narrowing at the suture line or flattening of the main pulmonary artery and its branches with concomitant reduction of its cross-sectional area caused by intensified forward tension at the site of the anastomosis, either because of inadequate growth of the main pulmonary artery [33] or because of the Lecompte procedure itself [19], are reported. Stenoses of the pulmonary artery branches, however, are considered as sequelae of supravalvular main pulmonary artery stenosis [22,33].

In conclusion, our study confirms good cardiological results of the neonatal ASO for TGA in patients with or without VSD. General health status is excellent in most children, while increased incidence of abnormal thoracic configuration after neonatal median sternotomy must be considered. Continuous long-term observations are necessary including ECG, Holter monitoring, exercise testing and careful echocardiographic evaluation in order to assess rhythm, function of the left systemic ventricle, development of the neo-aortic root and valve insufficiency as well as supravalvular pulmonary stenosis and growth of the pulmonary arteries.

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