The heart in Down syndrome
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WANTED! 8000 Heart Patients.
Identification of adult patients with a congenital heart defect lost to follow-up

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Nowadays, 85% of infants with congenital heart defects (CHD) reach adulthood and this adult population is expanding rapidly. Advances in cardiac surgery made operative repair of CHD possible. However, complete cure is seldom achieved. In most repaired defects late complications and residual lesions occur frequently. The misperception of "cure" has potentially serious consequences as patients do not seek continued medical advice and are lost to follow-up. Furthermore, young adolescents growing-up with CHD might struggle with psychosocial concerns because a complete recovery is impossible. These patients strive to lead a “normal” life. As a consequence patients deny their chronic disease and do not seek medical care. Probably a large proportion of adults with CHD fails to receive regular cardiac care and is lost to follow-up. A recent study reported that 61% of adult patients with congenital heart disease (CHD) fail to receive cardiac follow-up in Canada. In the Netherlands, we estimated that one third (approximately 8000) of adults with CHD are lost to follow-up (see method section). Although the estimated number of patients who fail to receive follow-up is lower in the Netherlands, we still believe it is unacceptably high. To increase awareness of adult patients with CHD and to identify patients who are lost to follow-up, the CONCOR project group initiated a nationwide media campaign entitled, "Wanted! 8000 Heart Patients".

In January 2009, we started a national campaign to identify adult patients with CHD lost to follow-up. The campaign consisted of a nationwide call via billboards and advertisements. In total approximately 1200 billboards were placed in 24 cities nationwide for a period of 1 to 2 weeks from January 2009 to May 2009. Adults with CHD between 20 to 40 years old, who were lost to cardiac follow up, were incited to call the CONCOR project group. This age-group was specifically mentioned to avoid confusion with acquired cardiac lesions.

During the campaign and thereafter, adults with CHD were able to call the CONCOR project group or could register themselves via a website, specifically designed for this campaign (www.8000vermisten.nl.) Three research nurses specialized into CHD, handled incoming patient calls and emails. After registration, patients were sent a CONCOR patient registration form and were advised to contact their general practitioner and to make an appointment with a cardiologist. After one year, patients were contacted again to collect information about the follow-up after
registration. Data on how many patients were seen by their cardiologist and the clinical consequences and events were assessed.

From January 2009 till January 2010, 1261 patients have contacted the CONCOR project group by telephone and email. In total, 593 patients (65% female, mean age 40 ± 12 years) were included. Table 1 shows all included CHD, of which 1.0% was severe CHD. After 1 year, 80% of the patients have been called again by a research nurse. In total, 66% of patients had contacted their cardiologist. Twenty-two percent required cardiac follow-up within 1 year. In 53 patients (16%) cardiac examination resulted in 76 new findings: 20% related to the aortic valve and aorta, 14% to the pulmonary valve, 10% to the mitral valve, 18% were arrhythmias or other electrical conduction abnormalities and a heterogeneous group (33%) of other symptoms (e.g. systemic hypertension, heart failure, tricuspid valve regurgitation). Most findings were mild, although 3 patients with repaired tetralogy of Fallot even needed pulmonary valve replacement shortly after registration.

Table 1 Congenital heart defects (n=593)

<table>
<thead>
<tr>
<th>Mild congenital heart defects, n (%)</th>
<th>504 (85%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial septal defect</td>
<td>182</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>176</td>
</tr>
<tr>
<td>Patent arterial duct</td>
<td>87</td>
</tr>
<tr>
<td>Pulmonary valvar stenosis</td>
<td>32</td>
</tr>
<tr>
<td>Other mild defects</td>
<td>27</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Moderate congenital heart defects, n (%)</th>
<th>83 (14%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic coarctation</td>
<td>46</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>13</td>
</tr>
<tr>
<td>Atrioventricular septal defect</td>
<td>11</td>
</tr>
<tr>
<td>Partially anomalous pulmonary venous connections</td>
<td>6</td>
</tr>
<tr>
<td>Sinus venosus atrial septal defect</td>
<td>5</td>
</tr>
<tr>
<td>Other moderate defects</td>
<td>2</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Severe congenital heart defects, n (%)</th>
<th>6 (1%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complete transposition of the great arteries †</td>
<td>4</td>
</tr>
<tr>
<td>Congenitally corrected transposition of the great arteries</td>
<td>1</td>
</tr>
<tr>
<td>Double chambered right ventricle</td>
<td>1</td>
</tr>
</tbody>
</table>

Numbers present total of new findings.
† 2 Senning procedure, 1 arterial switch operation, 1 unknown.
Our campaign has led to the identification of 593 lost patients with CHD. To our knowledge, this is the first time that data of a comprehensive national campaign on this actual topic has been published. Interestingly, not only patients with minor CHD were identified, but also patients with severe CHD were found (double chambered right ventricle and transposition of the great arteries). A substantial number of unknown residual lesions have been identified and a minority needed prompt intervention. According to current guidelines these patients with severe CHD need regular cardiac follow-up, as we know that long-term morbidity is high.\textsuperscript{10} Patients with prior cardiac surgery often consider themselves “cured,” although the majority faces a lifetime of problems including arrhythmias, heart failure, hypertension, vascular complications and one or more re-operations. Even patients with repaired “simple” lesions such as an atrial septal defect may not have normal survival if they are repaired in adulthood. Despite successful surgery for aortic coarctation, the prevalence of hypertension in adults 10 to 20 years after repair is 20% to 40\%\textsuperscript{11} and they may have premature cardiovascular complications.\textsuperscript{5,12,13} Besides registration we advised patients to contact their general practitioner and we encouraged to visit a cardiologist. In 16% of patients, unknown residual lesions were found of whom 6% needed an operation. Extrapolation to the total group of patients lost to follow-up would indicate that a substantial number of patients have unknown residual lesions. A minority of these patients needs a prompt intervention.

Thirty-four percent of patients did not visit a cardiologist due to various reasons. A remarkable and common finding was that the general physician disagreed with the need to visit a cardiologist. This indicates that awareness among general practitioners on this topic is still limited. Unfortunately, for some patients, the financial contribution for a doctor’s visit was a reason to refrain from follow-up. Another interesting finding was that 65% of the included patients were female, in contrast to the 50% female patients in the CONCOR registry. Apparently, the campaign has attracted more attention among women.

In conclusion, we were able to register a considerable amount of “missing” adults and the success of our campaign is a great stimulus to continue our efforts to increase awareness in the general population and retrieve those patients who are lost to follow-up. Furthermore our study can serve as a model for other countries to start similar programs.
In 2008, the Netherlands Society of Cardiology organized a sponsored bicycle ride from Amsterdam (the Netherlands) to the conference of the European Society of Cardiology in Munich (Germany). The raised 32000 euro was aimed for CONCOR as an unrestricted grant to realize the campaign. The authors of this manuscript have certified that they comply with the Principles of Ethical Publishing in the International Journal of Cardiology.\textsuperscript{14}

REFERENCES


