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Congenital heart defects are under-recognised in adult patients with Down's syndrome

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ABSTRACT

Background Congenital heart defects (CHD) are common in patients with Down's syndrome; however, patients living in residential centres have not always been screened for CHD in the past. The aim of this study was to investigate the prevalence of CHD in patients with Down's syndrome living in residential centres, and to determine whether cardiac screening should be recommended.

Methods Between January 2007 and November 2009 Dutch residential centres nationwide were randomly sampled. Medical files of all patients with Down's syndrome were investigated to retrieve documented information on known CHD. Echocardiography was performed on patients with unknown cardiac status. The main outcome measure was the number of newly diagnosed cases of CHD in adult patients with Down's syndrome.

Results Thirty-one centres and 1158 patients were included in the first stage of the study. Overall prevalence of known CHD was 16% (189 defects). Screening was performed in 138 patients without known CHD. In total, 24 new patients (17%) with a CHD were found, of which six patients needed semi-urgent care. Furthermore, 77% of the screened patients had mild to moderate regurgitation in one or more heart valves. Overall prevalence of CHD in adult Down's syndrome patients living in residential centres would be estimated at 33%.

Conclusions Seventeen per cent of patients with Down's syndrome living in residential centres had undiagnosed CHD, and valvular regurgitation was present in the majority of patients. Cardiac screening is recommended in older Down's syndrome patients, for whom new therapeutic options are available and for prevention of cardiac complications in old age.

INTRODUCTION

Congenital heart defects (CHD) are common in patients with Down's syndrome (DS). Approximately 50% of neonates with DS have a CHD. An atrioventricular septal defect (AVSD) is one of the most common defects, with a prevalence of 45% of cases, followed by a ventricular septal defect (VSD) in 35% and an atrial septal defect (ASD) in 10%.¹⁻⁴ Tetralogy of Fallot and a patent arterial duct (PDA) account for 10% of the cardiac defects in DS, whereas other defects are rare. Life expectancy of children with DS has been substantially increased in recent decades, mainly due to improvements in neonatal care and successful early cardiac surgery.^{5,6} Nowadays in most developed countries, all newborns with DS are evaluated for CHD by a paediatric cardiologist, and, in case of a defect,

followed by early corrective surgery, but this was not standard care before the 1980s.⁷ Echocardiograms are unlikely to have been performed on a large group of adult DS patients living in residential centres, thus it is very likely that underdiagnosis of CHD exists in this population. In patients with DS, late complications of uncorrected CHD, such as pulmonary hypertension or valve regurgitation, may occur, and the incidence increases with advanced age.^{8,9} Therefore, a large group of particularly 'older' patients without early surgical repair are at risk of having complications of their CHD. Identifying these patients and improving registration is necessary. Therefore, the aim of this study was to investigate the prevalence of heart defects in adult patients with DS who live in residential centres, and to investigate if cardiac screening for this population should be recommended.

METHODS

Study design

There are approximately 150 residential centres for people with intellectual disabilities in the Netherlands. This study consisted of a two-stage investigation (figure 1). First, residential centres nationwide were randomly sampled to participate in a cross-sectional study. Medical files of adult patients with DS were investigated to retrieve information on cardiac status, cardiac interventions and baseline patient characteristics. The second stage was a prospective cardiac screening programme, in which residential centres and their patients were asked to participate. Patients were eligible for participation when no information or doubtful information on cardiac status was retrieved from their medical files. Patients who were known to have a CHD, who were already being treated by a cardiologist and patients with severe Alzheimer's disease were excluded from cardiac screening. All included patients underwent physical examination and had an echocardiogram performed by an experienced ultrasound technician and evaluated by a cardiologist. All echocardiograms were performed with a portable General Electric VIVID I (Horten, Norway). Images were acquired and recorded digitally, and analysed offline. Apical, parasternal and subcostal views were obtained according to recommendations of the American Society of Echocardiography.¹⁰ Patients were screened for the existence of structural cardiac defects and a qualitative assessment of valvular regurgitation was performed. Approval was obtained from ethical boards of all participating institutions and informed consent was acquired from all subjects and/or their legal guardians.

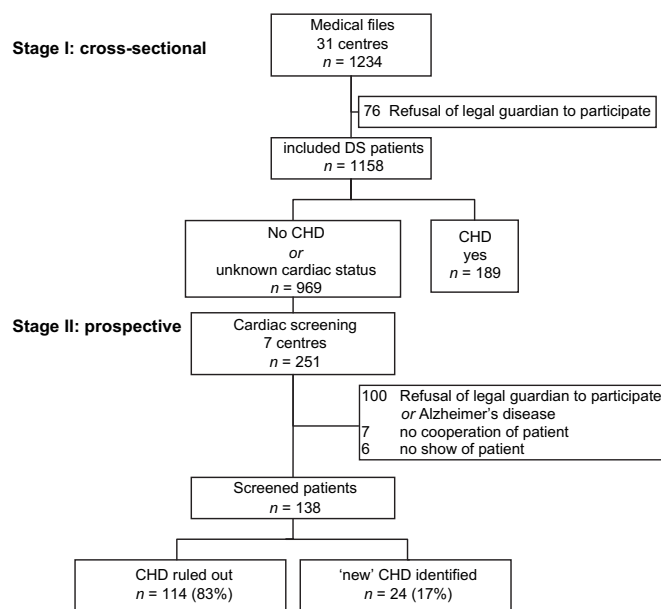


Figure 1 Study design of patient inclusion, DS, Down's syndrome; CHD, congenital heart defect.

Statistical analysis

Descriptive statistics were used to describe patient characteristics and type of heart defects. Differences between groups were analysed by unpaired Student t test for continuous variables and χ^2 test for nominal variables. Data are displayed as mean \pm SD, and the level of significance was set at $p < 0.05$. Statistical analysis was performed with SPSS 15.0.

RESULTS

Stage 1: outcome medical files

Thirty-one centres with a total of 1234 patients with DS were included in this study. The mean number of DS patients per centre was 37 (range 1–160). Seventy-six patients were excluded, because informed consent could not be obtained from the patient's legal guardian. Chart reviews were performed on 1158 included patients (mean age 46 ± 11 years; 54% males). Fourteen patients (1.2%) were younger than 18 years of age. Table 1 shows baseline patient characteristics. Overall prevalence

Table 1 Baseline characteristics (stage 1)

Characteristics	Total (n = 1158)
Age, mean (SD), years	46.5 (11.1)
Male sex, no. (%)	625 (54)
Level of intellectual disability (%), n=528	
mild	4
moderate	46
severe	40
profound	10
Patients with CHD, no. (%)	189 (16)
Patients without CHD, no (%)	680 (59)
Patients with unknown cardiac status, no. (%)	289 (25)
Patients with cardiac surgery, no. (%)*	43 (4)
PAH or Eisenmenger's syndrome, no. (%)	33 (3)

*Five patients had cardiac surgery for indications other than CHD, for example pericarditis, arteria lusoria.

CHD, congenital heart defect; PAH, pulmonary arterial hypertension.

of known CHD was 16% of patients (n=189; 14% men and 17% women). In 25% of the cases, the presence of a CHD could not be confirmed or rejected, because information on cardiac status was missing, or a CHD was suspected but not confirmed by cardiac imaging or other diagnostic tools. Table 2 shows the proportion of the various heart defects in subjects with known CHD. In patients with a known CHD, no difference in the prevalence of ASD, VSD and AVSD could be found between men and women. However, significantly more women with a CHD had a PDA compared to men (10 versus three respectively, $p=0.05$). Corrective cardiac surgery was performed in 20% of the patients with a CHD.

Stage 2: cardiac screening programme

Congenital heart defects

Seven of the 31 residential centres in the original sample participated in the cardiac screening program (figure 1). These seven centres having more than 2800 clients with an intellectual disability and 251 eligible DS patients were randomly sampled. After exclusion of 114 patients who had Alzheimer's disease, did not consent to undergo examination or refusal by the patient's legal guardian, cardiac screening was performed in 138 patients (see table 3 for baseline characteristics). In total 24 new patients were found in this population (prevalence of 'newly' diagnosed CHD 17%), varying between mild defects, such as mitral valve

Table 2 Congenital heart defects found by medical chart review of 1158 patients (stage 1)

Type of CHD	Number of defects (n = 207)	Men (n = 89)	Women (n = 91)	Prevalence CHD on total group (%)	Prevalence within CHD group (%)
VSD	65	28	35	5.6	31.4
AVSD	36	18	17	3.1	17.4
ASD	36	17	15	3.1	17.4
PDA	13	3	10	1.1	6.3
Other	57	31	22	5.0	27.5
pulmonary stenosis	13	8	4	1.1	6.3
mitral valve prolapse	11	7	4	<1	5.3
Tetralogy of Fallot	2	2	0	<1	<1
DCRV	2	1	1	<1	<1
cleft mitral valve	2	1	1	<1	<1
aortic stenosis	8	4	4	<1	3.9
aortic coarctation	2	1	1	<1	<1
mitral stenosis	2	2	0	<1	<1
bicuspid aortic valve	3	0	3	<1	1.4
unknown	12			1.0	5.8

ASD, atrial septal defect; AVSD, atrioventricular septal defect; CHD, congenital heart defect; DCRV, double-chambered right ventricle; PDA, patent arterial duct; VSD, ventricular septal defect.

Congenital heart disease

Table 3 Baseline characteristics of screened patients (stage 2)

Characteristics	Total (n=138)	Patients with CHD (n=24)	Patients without CHD (n=114)	p
Age, mean (SD), years	44.4 (10.9)	47.0 (9.6)	43.8 (11.1)	0.20
Male sex, no. (%)	62 (45)	63	41	0.07
Level of intellectual disability (%)				
mild	2	0	3	0.27*
moderate	58	68	56	
severe	32	18	35	
profound	8	14	6	

*Comparison of the distribution across the four categories of level of intellectual disability, based on Pearson χ^2 test
CHD, congenital heart defect.

prolapse without mitral valve regurgitation, which could be followed conservatively, and defects that needed semi-urgent care or evaluation by a cardiologist. The diagnosed cardiac defects in 24 screened positive patients are listed in table 4. Age, sex and severity of intellectual disability were not significantly different between screened positive and screened negative patients. All patients with a newly diagnosed CHD, except one, were born before 1980. In all centres, screened positive patients were found, varying from one to six patients.

Valvular regurgitation

The prevalences of valvular regurgitation in 138 patients are shown in figure 2. In 77% of the screened patients, mild to moderate regurgitation was present in one or more valves. Severity of valvular regurgitation was not correlated with age and sex. Mild to moderate regurgitation of the aortic valve was present in 38 patients (mean age 46 years, range 23–66 years), of which five had aortic stenosis (median age 51 years, range 42–66 years). The remaining 32 patients had isolated mild to moderate aortic valve regurgitation without aortic dilatation, leaflet perforation or calcifications.

DISCUSSION

This is the first study to investigate cardiac defects in a large group of adult patients with DS living in residential centres. The main finding is the underdiagnosis of cardiac defects in these patients. Although a mere 16% of patients had previously been diagnosed with congenital heart disease, it was found that a total of 33% of patients with DS living in residential centres had congenital heart disease. In accordance with previous

studies, the majority of these patients had septal defects.^{1 11} Prevalence studies on CHD in patients with DS have reported 50% of live born patients to have CHD. The present study revealed a lower number, which is most probably due to the adult study population. The patients in the present study are 'survivors' of a DS population born in the 1960s and 1970s, of which many patients with (severe) CHD had already died at a younger age. To obtain information on the prevalence of CHD in DS patients who had not been previously screened, cardiac screening was performed in 138 patients in whom information on CHD was missing or inconclusive. It was found that 24 (17%) of these patients had CHD, of whom 18 patients could be treated conservatively and six patients needed semi-urgent care or cardiac evaluation by a cardiologist.

In addition to CHD, a high prevalence of valvular regurgitation was found. The screening programme identified 38 patients with mild to moderate aortic regurgitation who should be followed-up by a cardiologist on a yearly basis, and in whom echocardiography should be performed every 2 years according to the guidelines of the European Society of Cardiology.¹² None of these patients needed immediate medical intervention. According to the Carpentier classification of aortic regurgitation, the majority of these patients (87%) could be classified as type II (without aortic dilatation, leaflet perforation or significant calcifications).¹³ A 10–30% prevalence of valvular regurgitation in young adults with DS was previously reported in small studies,^{14–16} and may be associated with collagen abnormalities in DS. As specific genes on chromosome 21 are overexpressed in patients with DS, collagen expression could be different in these patients. It is known that in patients with Down's syndrome the ligaments that normally hold the joints stable can be very slack. This can lead to an unusually wide range of movement at some joints, such as the atlanto-axial joint. Hypothetically, this abnormal collagen expression also results in valvular dysfunction in the ageing adult with DS. In this respect, type collagen VI is of particular interest because genes encoding the $\alpha 1$ and $\alpha 2$ chains are located on chromosome 21 and collagen VI is more prominent in fetal trisomy 21 hearts than in normal hearts.¹⁷

The main question remains of the number of DS patients that are potentially at risk. With a total of approximately 35 000 individuals with an intellectual disability living in residential centres,¹⁸ and a 10% (range 7–11%) prevalence of DS in participating centres, it is estimated that approximately 3500 adult patients with DS live in residential centres in the Netherlands. This means that 600 adult patients with DS who

Table 4 Outcome of cardiac screening (stage 2)

Newly diagnosed cardiac defects	n	Consequence
Mitral valve prolapse without mitral valve regurgitation	1	None
Bicuspid aortic valve without aortic valve regurgitation	1	Consultation by a cardiologist in 5 years
Mitral valve prolapse + tricuspid valve prolapse	1	Consultation by a cardiologist in 5 years
Mitral valve prolapse + mild mitral valve regurgitation	7	Consultation by a cardiologist in 5 years
Mitral valve prolapse + moderate mitral valve regurgitation	5	Consultation by a cardiologist in 3 years
Atrial septal defect + mitral valve prolapse	1	Consultation by a cardiologist in 3 years
Atrial septal defect	2	Consultation by a cardiologist in 3 years
Atrial septal defect + dilated and hypertrophied right ventricle	1	Referred to cardiologist for atrial septal defect closure; parents refrained from an intervention, yearly follow-up
Dysplastic mitral valve	1	Referred to cardiologist
Double-chambered right ventricle	1	Referred to cardiologist*
Partial atrioventricular septal defect + cleft mitral valve	1	Referred to cardiologist; surgical indication; however, no intervention due to comorbidity
Patent arterial duct	1	Referred to cardiologist for cardiac computer tomography; silent duct; no intervention
Cleft mitral valve + severe mitral valve regurgitation	1	Referred to cardiologist; cardiac follow-up yearly
Aortic valve regurgitation (mild-moderate)	38	Follow-up by echocardiography every 2 years

*Patient died suddenly before consultation by a cardiologist was done.

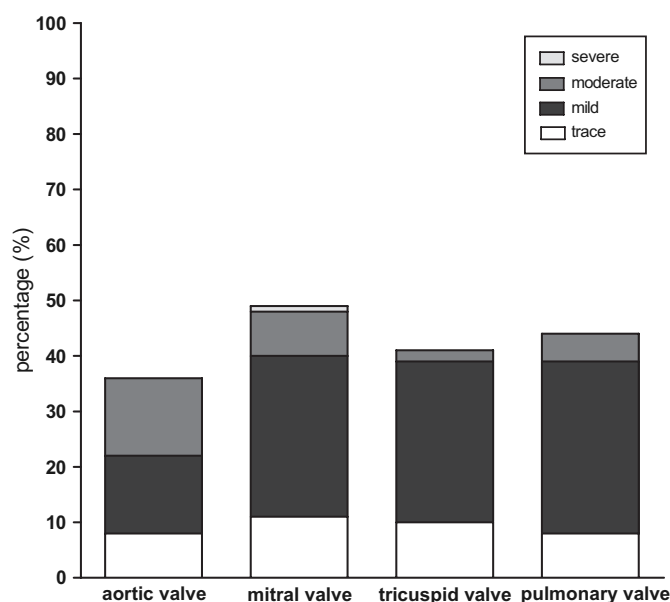


Figure 2 Valvular regurgitation in 138 screened patients.

live in residential centres have an undiagnosed CHD. In addition, an even larger group of almost 1000 patients have undiagnosed mild-to moderate aortic regurgitation. It is estimated that the overall prevalence of CHD in DS patients living in residential centres is 33%.

It seems highly likely that a similar underdiagnosis exists in other countries with residential services. However, many countries in Western Europe have experienced deinstitutionalisation, from large segregated and geographically isolated institutions to community-based residential facilities.¹⁹ Nonetheless, in many developed countries, residential facilities remain dominant, analogous to the Dutch situation.^{19 20} Although screening outside the residential setting was beyond the scope of this study, previous studies have reported hidden pathology found on adults with an intellectual disability living in the community.^{21 22} Underdiagnosis of CHD could also be present in adults with DS living in community-based residential centres. The present findings support cardiac screening of adults with DS as they fit badly into a system of healthcare delivery in which no care is received unless it is asked for.

Several strengths of this study are worthy of comment. Medical chart review was performed in a large group of individuals with DS nationwide. The screening programme was highly successful, with successful examination of 95% of the screened patients, due to the safe and familiar surroundings of the residential centre in which the screening was performed.

This study has some limitations. The first is the relatively small number of patients who agreed to participate in the second phase of cardiac screening in those without known cardiac defects. Population bias could be introduced, as only eight centres were contacted to participate in the screening programme, of which one decided not to participate. However, the participating centres are located in different regions of the country, varying in number of residing patients and all under care of specialised physicians for people with intellectual disabilities. Level of intellectual disability of the screened patients was according to the distribution in the other participating centres. Therefore, the seven centres are, for the greater part, representative for other residential centres in the Netherlands. Another concern is that a large proportion of patients could not participate in the screening programme,

because informed consent could not be obtained from their legal guardian. A single cardiac screening was performed and the long-term effect on morbidity and life-expectancy is not known. Further follow-up is needed to investigate the benefit of screening in this population.

CONCLUSION

In this study, 17% of adult patients with DS, who live in residential centres in the Netherlands, had undiagnosed congenital heart disease. Moreover, 77% of patients had unknown regurgitation of one or more heart valves. The authors advise large-scale screening of adult DS patients, to rule out congenital and valvular heart disease, and to provide these patients with adequate cardiac therapy.

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Competing interests R.B. reports having received an honorarium from Actelion Pharmaceuticals Nederland B.V for performing the echocardiograms.

Patient consent Obtained.

Ethics approval This study was conducted with the approval of the Ethical Boards of the participating residential settings and the Academic Medical Center.

Contributors All authors were involved in the conception design and data collection of the study and preparation of the manuscript, have read the manuscript and agree with the manuscript.

Provenance and peer review Not commissioned; externally peer reviewed.

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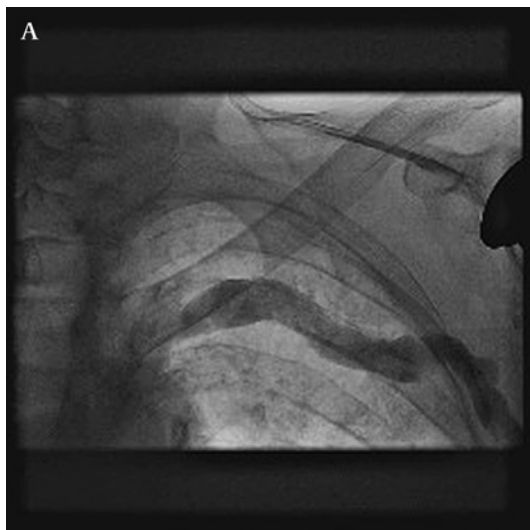
Images in cardiology

Central venous spasm during pacemaker insertion

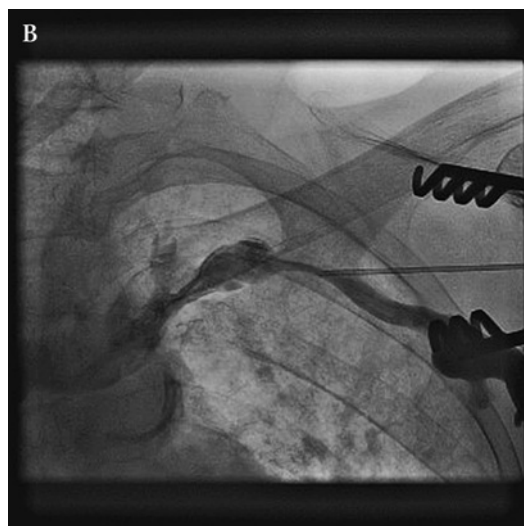
An 83-year-old female presented with syncope. She was found to be in complete heart block with an escape rhythm of 40 bpm. Following admission she remained haemodynamically stable with no prolonged pauses; she was therefore listed for pacemaker implantation the next morning.

Initial attempts at venous access were right-sided due to recent left shoulder surgery. A venogram revealed good calibre venous anatomy. Several attempts were made at venous puncture with good flashback, but attempts to pass standard and hydrophilic guidewires were unsuccessful. The patient became uncomfortable so the procedure was abandoned.

Left-sided access was attempted the next day. The patient was adequately hydrated and comfortable. An initial venogram showed reasonable calibre veins (see panel A), no cephalic vein was identified clinically or radiologically. Attempts at venous cannulation were again limited by an inability to advance a guidewire. A venogram was taken and revealed significant narrowing (see panel B), an 18G needle is also seen. Some time



Panel A



Panel B

was allowed to pass to allow the vessel to relax. We returned to puncture and were able to pass a hydrophilic wire into the axillary vein. A dual chamber pacing system was implanted. Follow-up the next day and at 6 weeks was unremarkable.

A detailed literature review revealed one previous similar case. Central venous spasm was associated with a catheter passing through the inferior vena cava (IVC) and required intravenous nitrate to relieve spasm.¹ This is the first case reported to resolve without nitrates, and the first to be published with cineradiography.

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Competing interests None.

Patient consent Obtained.

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