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Tropical Doctor

Tropical Doctor
0(0) 1–5

DOI: 10.1177/0049475518788468
journals.sagepub.com/home/tdo
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Adherence to health management guidelines for children with Down's syndrome at the Bustamante Hospital for Children, Jamaica

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Abstract

Health surveillance of children with Down's syndrome may be inadequate. We aimed to assess adherence to health management guidelines at the main paediatric hospital in Jamaica. Ours was a retrospective descriptive study over a five-year period. Data on demographics, co-morbidities, investigations, referrals and interventions were recorded. Of 41 children included in the study, 85% were diagnosed in the neonatal period. Congenital heart disease in 29 (76%) and ophthalmological disorders in 13/24 (54%) were the most common co-morbidities. Evaluations in accordance with the American Academy of Pediatrics guidelines were carried out in only 46% of the children for echocardiography, 48% for ophthalmology, 30% for hearing evaluation and 10% for neonatal thyroid screening. Thus, the recommended guidelines were not carried out in a timely manner in the majority of the children. Education of healthcare providers and caregivers along with the provision of adequate resources may help to resolve this inadequacy.

Keywords

Down's syndrome, trisomy 21, health surveillance, health maintenance

Introduction

Children with Down's syndrome (DS) can now be expected to reach the sixth decade of life. This is owing mainly to improvements in health management and the outcome of early cardiac surgery.[1] Early detection of and intervention for co-morbidities in affected children is important as this may significantly affect level of functioning.[2] Healthcare for patients with DS may be provided by a family physician or a general paediatrician, be decentralised or occur at a multidisciplinary facility.[1,3,4]

Guidelines have been developed to detect and monitor co-morbidities in patients with DS. The American Academy of Pediatrics (AAP) published its latest guidelines in 2011. These are organised into clinical features and problem issues that need to be addressed from the prenatal period to adolescence.[3] The Down's Syndrome Medical Interest Group UK and Ireland (DSMIG) published the latest revision of its guidelines in 2012. These are provided for evaluation of cardiac disease, thyroid conditions, hearing deficiencies, ophthalmic complications and suitable monitoring of growth.[5] Other countries such as Finland have also published their own national guidelines.[4]

There have been recent studies that have reviewed the adherence to management guidelines and adequacy of health management of patients with DS. The degree of adherence varies from country to country. A review of the adequacy of medical follow-up in Israel showed there were deficiencies in the evaluation of thyroid function as well as vision and hearing and cervical spine stability.[6] A retrospective observational study in Sri Lanka reported that the mean age of initial

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assessments for cardiac, hearing, vision and thyroid function was delayed.[7] In Canada, in all age groups, only 50% of patients reported medical referrals or laboratory tests.[8]

No previous study has been conducted in Jamaica.

Methods

The Bustamante Hospital for Children (BHC) is the first specialist hospital for children established in the English-speaking Caribbean and manages patients from birth to 12 years. It has a bed capacity of 283, inclusive of a five-bed intensive care unit. The Outpatient Department conducts general and subspecialty clinics, the latter including Cardiology, Ophthalmology, ENT, General Surgery and Orthopaedics. It is the main paediatric hospital in Jamaica and one of two hospitals that provides cardiac services for children, but it is the only one to do so free of cost. Therefore, the majority of children with DS receive cardiac screening at the BHC although follow-up care for these children may occur at other institutions.

Ours was a retrospective, descriptive study looking at all children aged 0–12 years with DS managed at the BHC between 1 January 2012 and 31 December 2015. Study participants were identified from the Patient Information Management System. All children with a diagnosis of DS or trisomy 21 (T21) admitted to BHC were included in the study. The medical records for all participants who fulfilled inclusion criteria were retrieved. A data extraction sheet was used to record data on demographics, co-morbidities, timing and results of investigations, timing of and compliance with referrals, and interventions.

Patients aged > 12 years and those with a diagnosis of DS or T21 referred from another institution for screening evaluation only and whose healthcare was not continued at BHC were excluded from this study.

The Early Stimulation Programme (ESP) is a programme implemented by the Jamaica Council for persons with disabilities that offers evaluation and early intervention for children with neurodevelopmental delay between birth and six years.[9]

The Jamaica Downs Syndrome Foundation (JDSF) is a non-profit organisation established in 2007. Its aim is to provide increased social acceptance of children with DS, provide parent education and promote early intervention.[10] In March 2013, the JDSF in collaboration with the Ministry of Health made the pneumococcal vaccine available free of cost to children with DS.

Descriptive analyses were performed. Statistical significance was taken at the level $P < 0.05$. Statistical analyses were performed using SPSS version 13 (SPSS Inc., Chicago, IL, USA).

Ethical approval

Ethical approval for the conduct of the study was granted by the Research Ethics Committees of the University of the West Indies, the Ministry of Health and the South Eastern Regional Health Authority.

Results

There were 98 children with DS or T21; a total of 36 followed up at another health institution or with a private physician were excluded. Six children who were aged >12 years and two children who defaulted from follow-up shortly after diagnosis were also excluded. There were 13 missing medical files, where it was impossible to determine which children had DS, as DS, T21 and dysmorphic syndrome have the same code in our patient information system. The medical outpatient logbooks do not list patients' diagnoses and so patients with T21 who were not admitted were not captured in this study. The total number of study participants was 41 (Figure 1), of whom 26 (63%) were boys. The age range of the children was 0.8–11.9 years with a mean of 5.6 ± 2.8. The age of diagnosis was documented for 33 children; of these, 28 (85%) were diagnosed in the neonatal period. One child was diagnosed after the age of three months. One participant had chromosomal studies done.

An echocardiogram was done in 40 patients (98%), of whom 25 (61%) had this first performed by the age of three months; from this group, 19 (76%) had the echocardiogram in the neonatal period (Figure 2). There was no documentation of age of first echocardiogram for three children. Echocardiography revealed 29 patients (76%) had a congenital heart lesion. The most frequent malformations found were atrial ventricular septal defect (AVSD) and patent ductus arteriosus (PDA) (Table 1). One patient who was referred for echocardiogram defaulted from care. Medical management was preferred for congenital heart disease (CHD) in 17 (53%), and only six (7%) had surgical intervention.

Ophthalmological and auditory referral was carried out in 29 patients (71%) (Figure 3), of whom 20 (69%) had this before their first birthday. Only 24 (83%) of the 29 children referred actually had their eye evaluation performed, and in 11 (46%) this was normal while the remainder had various abnormalities: strabismus in four (11%); nystagmus in three (13%); or some other abnormality in six (16%), including hyperopia and cataracts. Corrective eyewear was recommended for three children and surgery was performed on one child.

Auditory referral was carried out in 27 patients (66%) but a mere nine (31%) had an evaluation

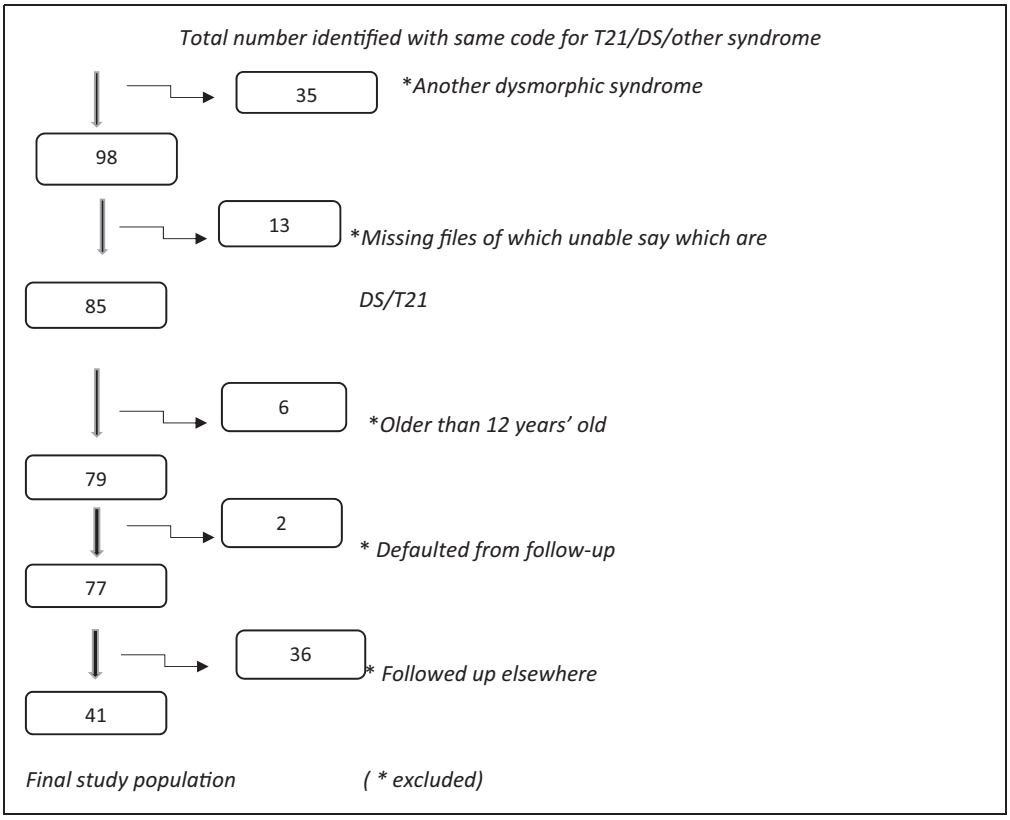

Figure 1. Flow chart indicating how final study population achieved.

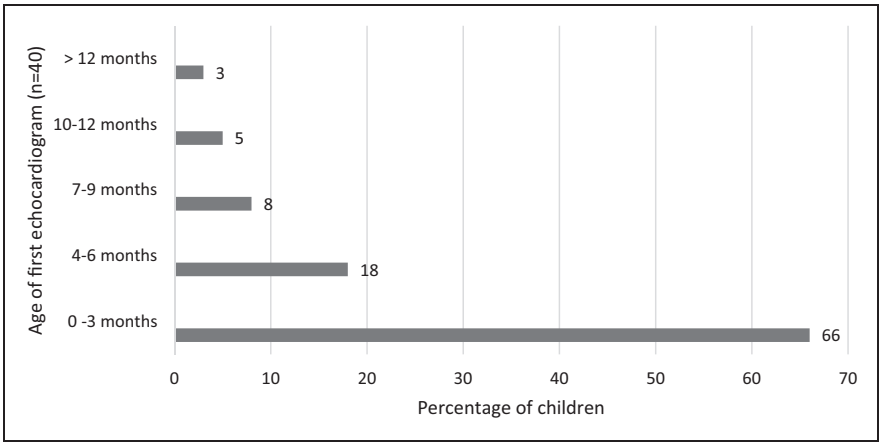

Figure 2. Age of first echocardiogram for children with DS followed up at BHS between January 2012 and December 2015.

done. One child had a hearing impairment, but no intervention was documented for this child. Thyroid function tests (TFTs) were performed in 27 children (71%) but none had hypothyroidism initially; 22 (82%) had repeat thyroid function tests performed

One child was referred for and received the pneumococcal vaccine. Some 22 patients (54%) were referred to ESP but only 50% attended. There was no documentation about formal assessment for cognitive impairment in any child. Obesity was reported in four (10%) and gastrointestinal atresia in a single child. Only seven children (17%) were referred to the JDSF. Three children died, all of whom had CHD. One patient died postoperatively for repair of his CHD, while the other two died of complications of heart failure and a lower respiratory tract infection.

Appropriate investigation and follow-up for cardiac, hearing, sight and thyroid function based on AAP guidelines was achieved in a minority of children (Table 2).

Discussion

Our study found that adherence to health management guidelines for children with DS at our hospital was suboptimal. The diagnosis was made clinically in almost all children and results of chromosomal studies were only documented for one child. This is in stark contrast to studies in high-income countries where 80–100% receive chromosomal analysis.[7,11,12] Reasons for this in our setting include lack of accessibility of the tests owing to cost and reluctance of parents to have the tests done, partly owing to advanced age of the parents and their absence of a desire for more children. Nonetheless, prior studies have confirmed the accuracy of a clinical diagnosis in children with DS.[11,12]

Since the majority of children in our study were referred to BHC from other institutions, we relied on parent recall to determine the age of first diagnosis thus introducing a recall bias. Almost 90% were diagnosed in the neonatal period but not necessarily in the first week of life. This has implications for certain screening tests such as thyroid function tests and echocardiograms that ideally should be done shortly after birth.[11,12]

CHD was common and almost half of the children had an echocardiogram performed before they were one month old, which is a vast improvement over the

Table 1. Types of CHD seen in children (n = 40) with DS followed up at BHC between Jan 2012 and Dec 2015.

Results of ECHO	n (%)
AVSD	12 (30)
PDA only	5 (12)
AVSD and PDA	8 (20)
VSD and PDA	3 (8)
TOF	2 (5)
Normal	8 (20)
Other	2 (5)
Total	40 (100)

Table 2. Number of children (n = 41) with DS followed up at BHC between January 2012 and December 2015, who had investigation in accordance with AAP guidelines.

Recommended guideline	n (%)
Echo at birth–1 month	21 (46)
Ophthalmology referral to assess for strabismus, cataracts and nystagmus at birth–1 year	20 (48)
TSH at birth–1 month	12 (30)
New-born hearing screen at birth–1 month	4 (10)

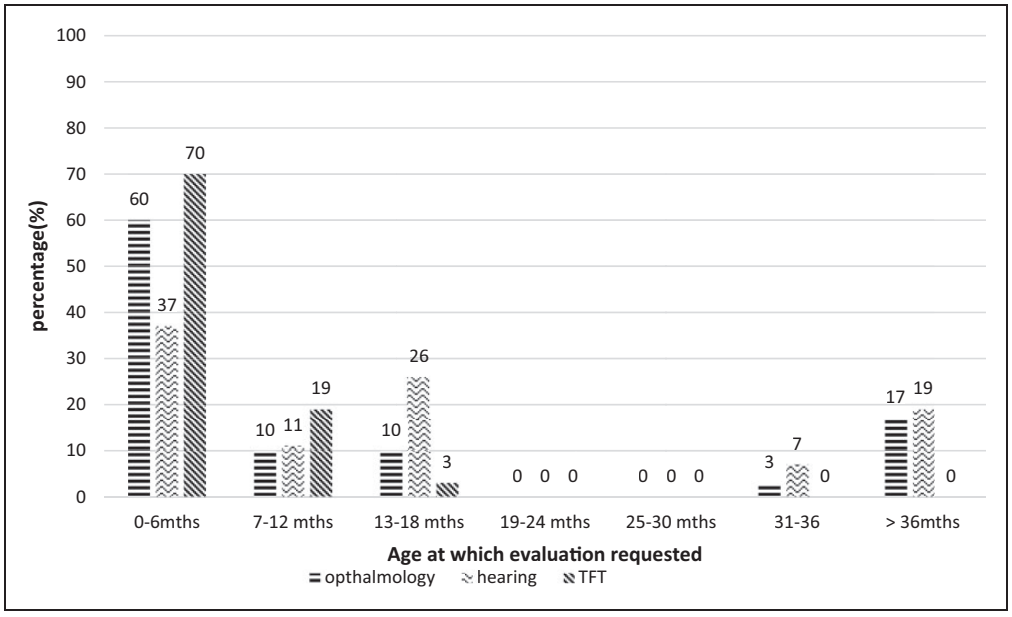

Figure 3. Comparison of the ages at which ophthalmology, hearing evaluations and thyroid functions were requested in children with DS followed up at BHC between January 2012 and December 2015.

9% previously documented in a 10-year retrospective Jamaican study in 2010.[14] Nonetheless, this is still sub-optimal.[6] BHC is the single public health facility in Jamaica which offers free echocardiograms for the paediatric population. Owing to high demand, delays are inevitable. By three months, however, two-thirds of the children in our study had received an echocardiogram.

The need for ophthalmological and auditory evaluation is evidently not well appreciated, although monitoring of thyroid function was good. It seems that physician awareness of the current arrangement between the JDSF and the MOH for free pneumococcal vaccination is wanting, as is the poor rate of referral to the JDSF for support. Similarly, despite no formal documentation of cognitive impairment, over half of the children were referred to ESP suggesting developmental delay was frequent. IQ testing or formal development assessment is not routinely performed.

Thus, the health management and follow-up of children with DS in Jamaica was inadequate and not in accordance with the AAP guidelines of 2011. A main contributing factor appears to be lack of knowledge of the healthcare provider and the caregiver, but also overloading of the healthcare system with other problems.

A limitation of our study is that patients with DS who were not admitted to hospital were not captured as their diagnoses were not documented in the clinic log books. Poor documentation also limited the data available.

As children with DS now have increased life expectancy, and with adequate management of co-morbidities and social support, their quality of life may be substantially improved; we urge education of healthcare providers and caregivers about associated co-morbidities and the available guidelines in place to monitor them. We recommend that a schedule of investigations, specialised growth charts and follow-up care for children with DS should be included in their health passport. An increase in the availability of and access to the requisite services within the public health service is mandatory and specialised clinics geared toward individuals with DS should be put in place.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) disclosed receipt of the following financial support for the research, authorship, and/or publication of this article: This research was self funded by the researchers.

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