Adherence to preventive health care in children and young adults with Down syndrome (DS) in Sri Lanka

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Abstract

Objectives To assess adherence to preventive care and age-appropriate screening in different age categories of Down syndrome (DS) persons.

Methods This community-based retrospective observational study on ambulatory persons with DS from different regions of Sri Lanka was carried out on 100 randomly selected individuals from a sample framework of approximately 300. Their screening behavior for cardiac, ophthalmic, hearing and thyroid status was evaluated. Age at first medical consultation for DS specific complications, whether undertaken when symptomatic or asymptomatic, and use of DS-specific growth charts were extracted from medical records. Screening was compared in three age groups (<10, 10-15, and >15 years).

Results One hundred clinically diagnosed DS individuals (F: M 1:1.2) aged 2-28 years (mean 13.5 years) were from varied socio-economic backgrounds and rural (68%) suburban (20%) and urban (12%) settings. They all had had postnatal medical assessments. Initial eye, ENT, cardiac consultations was at significantly earlier ages in those below 10 years. Age-appropriate cardiac assessments occurred in 58%, 25% and 7.5% in each age group. Vision, hearing and newborn thyroid status were not routinely screened for. Only 7% had special growth charts. Area of residence had no effect on screening behavior.

Conclusions Screening for complications in DS was delayed and inconsistent but occurred at significantly earlier ages in younger children. Use of appropriate growth charts was very low. Screening behavior was unaffected by area of residence. We recommend incorporating DS-specific medical checklists as a preventive health routine for this unique group of children.

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Introduction

Survival of children with Down syndrome (DS) has improved around the world due to early detection of this syndrome's numerous multi-organ complications (1-4). Timely preventive health screening protocols are accepted health policy in many countries (5-6). Although preventive medical checklists with local adaptations, were advocated they are still not widely used in Sri Lanka (7). The objective of this study was to assess adherence to preventive health care in a sample of ambulatory DS persons and compare screening behaviour in different age categories.

Methods

This community-based retrospective observational study, was carried out on 100 ambulatory children and young persons (2-28 years) with DS, randomly selected from approximately 300 DS individuals attending a free health camp on Down Syndrome Day 2013 in Colombo. Due to logistical constraints, sample size was limited to 100 persons who provided consent from accompanying adult/s and had comprehensive medical records available for scrutiny. Absence of written medical records was an exclusion criterion.

An interviewer-administered questionnaire recorded age, gender, places of birth and residence, dates of first and subsequent medical consultations for cardiac, ophthalmic, hearing and thyroid assessments. Whether symptomatic or asymptomatic at first consultations, identified co-morbidities and availability of a special growth chart were obtained from medical records. Patient demographics and screening behaviour were compared between the three age groups (2-<10 years, 10-15 years and > 15 years) using Z test for two sample means/ proportions.

Results

There were 100 clinically diagnosed DS individuals (44 females 56 males, F: M ratio 1:1.2) aged 2-28 years (mean 13.5 years) of varied socio-economic backgrounds. Area of residence were rural (68) suburban (20) urban (12). Entire sample was hospital-born and had postnatal medical examinations.

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Timing of investigations for DS specific complications and identified co-morbidities are given in Table 1. Cardiac assessment including of echocardiography was done in 66%. Age at first echocardiogram ranged from neonate to 17 years. Cardiac assessment was significantly earlier in younger age groups ($p< 0.05$) (Table 2) but only 29% had echocardiography by the recommended age of three months. Those < 10 years had highest coverage (58%) but was a preventive measure only in a minority (32%). Others were assessed because of "heart murmur" (61%), "breathlessness" (5%), and "recurrent chest infections" (3%). Heart defects were documented in 43 persons.

Ophthalmic assessment was done in 60% of whom 58% were asymptomatic. Squint, nystagmus and naso-lacrimal duct obstruction were the reasons for seeking ophthalmic services. First assessment was carried out by six months of age in only 7% and statistically significant ($p <0.05$) were seen between age groups (Table 2). In 35% ophthalmic assessment was after age 10 years. Only 48% had ENT assessments and of whom 69% had hearing assessed. Severe deafness was reported in 9%. Main reasons for ENT consultations were infections. Only 8% were assessed in infancy. Ophthalmic or audiological assessments did not take place annually. Thirty seven underwent thyroid hormone assay and 13 (35%) were on treatment for hypothyroidism.

Screening behavior in the three age groups found significant differences ($p<0.05$) in age of initial cardiac, eye and hearing assessments; and screening rates were highest in the youngest age group (Table 2). Area of residence showed no relationship ($p>0.05$) to screening behavior. Special DS-growth charts were used in 7%.

Table 1. Screening behaviour and co-morbidities in 100 Down Syndrome individuals aged 2-28 years

Screening behaviour	*Number (%)*
Echocardiography performed at least once (n=100)	66 (66%)
Echocardiography by age 3 months (n=100)	29 (29%)
Mean age of first echocardiogram (n=66)	3.2 years
Reason for echocardiography (n=66)	
Routine screening when asymptomatic	21 (31.8%)
Cardiac murmur	40 (60.6%)
Breathlessness	3 (4.5%)
Recurrent chest infections	2 (3.0%)
Cyanosis	0 (0%)
Heart defects identified (n=66)	43 (65%)
Type of heart lesion/s (n=43)	
ASD	16 (37%)
VSD	13 (30%)
PDA	8 (19%)
AV canal defects	3 (7%)
Complex cardiac lesions	12 (28%)
Heart surgery needed	22 (51.2%)
At least one eye assessment performed (n=100)	60 (60%)
Mean age of initial eye assessment (n=60)	9.0 years (SD=7.5 years)
Eye assessment by age 6 months (n=60)	4 (7%)
Reason for eye assessment (n=60)	
screening when asymptomatic	35 (62.5%)
squint, nystagmus, blocked naso-lacrimal duct *etc*	21 (37.5%)
Reason for eye check by six months infancy (n=4)	
abnormality detected by paediatrician	4 (100%)
Age at initial eye assessment	
Infancy	4 (6.7%)
1-5 years	20 (33.3%)
5-10 years	15 (25%)
> 10 years	21 (35%)
Number identified with eye abnormalities (n=60)	37 (61.7%)
Needed spectacles	27 (45%)
Needed surgery	2 (3.3%)
Annual check of vision	0
Number who sought ENT services (n=100)	48 (48%)
ENT assessment by six months (n=100)	4 (4%)
Number who had at least one hearing assessment (n=48)	33 (68.8%)
Severe deafness	3 (9.1%)
Age at initial ENT assessment (n= 48)	
Infancy	4 (8.3%)
1-5 years	22 (45.8%)
5-10 years	11 (22.9%)
>10 years	11 (22.9%)
Mean age of hearing assessment (n=33)	6.7 years (SD= 6.2 years)
Audiology annually	0
Thyroid status assessed (n= 100)	37 (37%)
Neonatal screening (n=37)	0
Hypothyroidism identified (n=37)	13 (35.1%)

Table 2. Comparison of screening / medical checks in three age groups in Down syndrome

	=/<10 years (n=33)	*>10-15 years (n=28)*	*>15 years (n=38)*
Mean age of initial echocardiogram	8.2 months	35.8 months	76.6 months
Echocardiography by 3 months of age	19 (57.6%)	7 (25%)	3 (7.9%)
Mean age at initial eye assessment	3.2 Y	6.67 Y	14.96 Y
Initial eye check at/before 6 months	3 (9.1%)	0	1 (2.63%)
Mean age of initial ENT encounter	2.33 Y	5.54 Y	12.53 Y
Initial ENT consultation by 6 months	4 (12.1%)	0	0
Neonatal thyroid screen	0	0	0
TH assay during infancy	16 (48.5%)	13 (46.4%)	7 (18.4%)

Discussion

In the absence of a national registry for DS in Sri Lanka, we used a registry of 350 DS persons, maintained by a national level non-governmental organization. Our sample framework consisted of approximately 300 DS persons of this registry who attended a health camp. Lack of medical records was not a significant problem in this sample. Preventive health checks on four organs that commonly affect quality of life in DS (heart, eye, ear and thyroid) were retrospectively investigated for, in this sample of 100 randomly selected DS individuals.

We found low screening rates and a substantial proportion without cardiac (34%), ophthalmic (40%) or hearing (67%) assessments. Annual ophthalmic and audiology screening which is invaluable during school years had not taken place at all. Even in the youngest age group which had the best cardiac screening rates, initial assessment was delayed in 42%. DS-specific growth charts were used only in a small minority. Access to medical facilities did not appear to be a contributing factor because there was no difference between rural and urban children.

Screening rates in DS persons not included in a registry may be even lower than those found in this study. We recommend a special preventive health program for DS children, incorporating DS-specific screening checklists and growth charts, to improve their health status and quality of life.

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