

# Neurodevelopmental evaluation and referral practices in children with congenital heart disease in central South Africa

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## INTRODUCTION

There is substantial evidence that congenital heart disease (CHD) survivors are at higher risk for developmental delays than the general population.<sup>(1-3)</sup> For this reason, neurodevelopment has become one of the most important outcomes being investigated in children with CHD.<sup>(1,2,4)</sup> As many as half of children with CHD present with neurodevelopmental delays characterised by mild impairments across multiple domains, including deficits in gross and fine motor skills, cognition and language, and inattention, impulsivity, impaired executive function and psychosocial problems.<sup>(5-8)</sup> For many parents, over time, the neurodevelopmental difficulties faced by their child outweigh the daily burden caused by their heart disease.<sup>(9)</sup> Developmental delays also have a high cost to society, with costs escalating considerably when the need for developmental intervention is not met and developmental delays are not addressed effectively.<sup>(10)</sup>

The American Heart Association (AHA) published a guideline targeting the evaluation and management of neurodevelopment in the CHD population in 2012, in an attempt to address

## ABSTRACT

**Introduction:** Children with congenital heart disease (CHD) are at higher risk for developmental delays than the general population. The American Heart Association (AHA) published a guideline to address these concerns in 2012. This study determined the neurodevelopmental evaluation and referral practices of practitioners in central South Africa.

**Method:** An online survey was administered to practitioners (n=45) including paediatric cardiologists (n=4), cardiothoracic surgeons (n=4) and general paediatricians (n=37). Information on practitioner characteristics, awareness of the 2012 AHA guideline; and neurodevelopmental evaluation and referral practices was collected.

**Results:** Twenty-one practitioners responded, including paediatric cardiologists (n=4), cardiothoracic surgeons (n=2) and paediatricians (n=15). Data for 20 practitioners was included. Despite most practitioners (n=18) indicating guidelines for the management of development were important, the majority (n=16; 80%) were unaware of the guideline. Most practitioners (n=18; 90%) failed to risk stratify children to identify those to be evaluated. Children with developmental delays were referred for formal developmental evaluation (n=11; 55%) and to intervention therapies (n= 15; 75%).

**Conclusion:** Most practitioners are unaware of the 2012 AHA guideline. Awareness of the developmental risks associated with CHD and implementation of the guideline could promote early identification of developmental delays with referral to intervention therapies.

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these concerns.<sup>(1,2,10)</sup> The guideline is not context-specific and serves as a universal tool to facilitate clinical decision-making based on best practice evidence.<sup>(4,11)</sup> The guideline uses a risk-stratification approach with a clinical algorithm, including a combination of developmental surveillance, screening and evaluation aimed at the early identification of developmental delays.<sup>(1,2,4,10)</sup> By extension, the successful implementation of the guideline also results in the appropriate referral of children with developmental delays to intervention therapies, including physiotherapy, occupational therapy and speech therapy, as well as for psychological and educational support – to optimise their developmental outcome.<sup>(1,12-14)</sup>

The awareness of the paediatric cardiac care community in South Africa (SA) of the 2012 AHA guidelines is unknown. This study aimed to determine the neurodevelopmental evaluation and referral practices of practitioners treating children with CHD in central SA, as well as practitioners' awareness of the 2012 AHA guideline.

## MATERIALS AND METHODS

Information on practitioner characteristics, awareness of the 2012 AHA guideline, and neurodevelopmental evaluation and referral practices were collected using a questionnaire. The questionnaire used in a survey to determine the implementation of the 2012 AHA guideline by cardiac care practitioners in the United States (US), was adapted and expanded.<sup>(2,4)</sup> Response options to questions on clinical practice settings were adapted to the South African context.

A question was added under the developmental screening section to explore practitioners' use of web-based applications for developmental screening. It was deemed important to explore this as a possible means of providing remote developmental care. Three further questions were added under the referral section to explore practitioners' views on the accessibility and barriers to intervention therapies. The content validity of the questionnaire was established by a physiotherapist familiar with the 2012 AHA guideline and experienced in the assessment of neurodevelopment in children with CHD in the South African context.

Forty-five practitioners, including general paediatricians (n=38), paediatric cardiologists (n=4) and cardiothoracic surgeons (n=4), practising in the Free State and/or Northern Cape provinces of SA were recruited into this observational descriptive study. No sampling method was used due to the small population. Practitioners were excluded if they did not treat children with CHD.

Ethical clearance was obtained from the Health Research Ethics Committee of the University of the Free State (UFS-HSD2017/1271). Participation was voluntary and completing and submitting the online survey provided informed consent. Confidentiality of participant information was maintained.

An introductory email was sent to 45 practitioners explaining the purpose of the survey, the informed consent procedure and the time required to complete the questionnaire. An open access copy of the 2012 AHA guideline was included in the email. The email contained a link to the questionnaire, with an access password. This was a closed survey, and was accessible

by email invitation only. The survey was created and managed by an online survey platform (EvaSys) hosted by the University of the Free State. Completed surveys were submitted electronically and participant responses were automatically captured in an EXCEL spreadsheet. Once participants submitted their questionnaire, the survey became inaccessible, preventing multiple submissions. The survey remained open for 6 weeks and email reminders were sent out to non-respondents.

Statistical analysis was done by the Department of Biostatistics at the University of the Free State. Descriptive statistics for the categorical participant responses are summarised using frequencies with percentages.

## RESULTS

Twenty-one practitioners responded to the survey (response rate of 46.7%). A practitioner (n=1) was excluded who did not treat children with CHD. The responses from 20 practitioners, including general paediatricians (n=14), paediatric cardiologists (n=4) and cardiothoracic surgeons (n=2), were included in the analysis. The highest response rate was from the paediatric cardiologists (100%), with the lowest response from the general paediatricians (36.8%). Most of the practitioners (n=13; 65%) had been practising in their specialised field for more than a decade. Most practitioners (n=17; 85%) practised in both the public and private healthcare setting. Half of the practitioners (n=10; 50%) had more than 25 children with CHD under their care, with the paediatric cardiologists (n=4, 100%) carrying the highest load – with more than 50 children under their care (Table I).

Most practitioners (n=18; 90%) felt that having neurodevelopmental evaluation and management guidelines for children with CHD was "somewhat" or "very important". Despite this view, the majority (n=16; 80%) were unaware of the 2012 AHA guideline. Awareness was lowest amongst general paediatricians (n=1; 7.1%). The vast majority of practitioners (n=18; 94.7%) did not risk stratify children with CHD in line with the 2012 AHA guideline (Table II).

Most practitioners (n=15; 75%) routinely performed developmental surveillance. In addition to their own surveillance, several practitioners (n=7; 46.7%) also referred children to an occupational therapist for developmental surveillance. Thirteen practitioners (65%) reported routinely referring children with CHD for developmental screening. Children were referred for screening most often at all care visits (n=15; 75%) and at 6 months of age (n=5; 25%). A single practitioner (5%) used electronic web-based applications for screening (Table II).

**TABLE I: Participant characteristics.**

| Variable   | Frequency (%)<br>(n=20) |
|--|-------------------------|
| <b>Medical speciality</b>  |                         |
| General paediatrician  | 14 (70%)                |
| Paediatric cardiologist  | 4 (20%)                 |
| Cardiothoracic surgeon   | 2 (10%)                 |
| <b>Survey response rate per medical speciality</b>                   |                         |
| Pharmacological concepts   | 14 out of 38<br>(36.8%) |
| Solutions: compositions and therapy                                  | 4 out of 4 (100%)       |
| Fluid balance and assessment   | 2 out of 4 (50%)        |
| <b>Years of clinical practice in current speciality</b>              |                         |
| <5 years   | 2 (10%)                 |
| 5-10 years   | 5 (25%)                 |
| 11-15 years  | 2 (10%)                 |
| >15 years  | 11 (55%)                |
| <b>Location of clinical practice*</b>                                |                         |
| Public health sector   | 3 (15%)                 |
| Academic or university-affiliated hospital                           | 5 (25%)                 |
| Public health sector and academic or university-affiliated hospital  | 3 (15%)                 |
| Private health sector  | 3 (15%)                 |
| Public and private health sector                                     | 2 (10%)                 |
| Public and private health sector; and university-affiliated hospital | 4 (20%)                 |

\*Multiple category responses permitted.

On the identification of developmental difficulties, practitioners most often referred children to allied health services (n=15; 75%) and medical specialists, including paediatric neurologists and neurodevelopmental paediatricians (n=11; 55%), for formal developmental evaluation. Less than a quarter (n=4; 20%) of practitioners referred children to an interdisciplinary clinic for developmental evaluation. Children were usually referred for formal developmental evaluation during infancy (n=14; 70%) and in the pre-school years (n=9; 45%). Referrals were usually based on the practitioner's own concerns about the child's development (n=17; 85%) and/or noted parental concerns (n=16; 80%). Most practitioners (n=15; 75%) referred children diagnosed with a genetic disorder for formal developmental evaluation (n=15; 75%). Only eight practitioners (40%) reported that their referrals for formal developmental evaluation were based on abnormal developmental screening test results (Table III). Practitioners identified the unavailability

of medical specialists including paediatric neurologists and neurodevelopmental paediatricians (n=8; 40%), and the distances families have to travel to access healthcare services (n=8; 40%) as the main barriers to formal developmental evaluation (Table IV).

When referring children to allied health services, practitioners usually referred children to occupational therapy (n=100%), followed by physiotherapy (n=15; 75%) and then speech therapy (n=14; 70%). Only three practitioners (15%) referred children to a psychologist. Practitioners felt that the unavailability of therapy services (n=15; 75%), the distances families have to travel to access therapy (n=15; 75%) and the time and financial cost to parents in having to take their children to therapy (n=8; 40%), were barriers to accessing intervention therapies (Table IV).

## DISCUSSION

Despite most practitioners indicating that having neurodevelopmental evaluation and management guidelines for children with CHD was important, most (80%) were unaware of the 2012 AHA guideline. Awareness was lowest among the general paediatricians (n=1; 7.1%). A similar lack of awareness of 79% was reported in the US survey.<sup>(4)</sup> Several factors could explain this lack of awareness. The guideline was published in a cardiovascular journal which may have meant it was less likely to be read by general paediatricians.<sup>(4)</sup> Paediatric cardiologists and cardiothoracic surgeons are primarily focused on the cardiovascular health of the child with CHD, and may be unaware of or overlook the importance of the child's neurodevelopmental outcome, as a measure of the success of their cardiac interventions.<sup>(2,8,15)</sup> The fact that the guideline is considered "American", despite its universal relevance, may also have deterred South African practitioners from reading it.

Paediatric cardiologists and cardiothoracic surgeons are primarily responsible for the care provided to children with CHD in South Africa. The shortage of paediatric cardiologists in the country makes it highly probable that many children with CHD are cared for by general paediatricians.<sup>(16,17)</sup> The poor response from general paediatricians (36.8%) may indicate that they are either not treating children with CHD and/or that they are unaware of the developmental risks faced by these children.

Only three-quarters of practitioners performed routine developmental surveillance at every visit to identify those children at risk for developmental delays.<sup>(11)</sup> Cardiothoracic surgeons performed no developmental surveillance. This is considered a missed opportunity, as they are ideally situated

TABLE 11: Survey response of practitioners (Part I Awareness of the 2012 AHA guideline and surveillance practices).

| Variable  | All (n=20) | Paediatricians (n=14) | Paediatric cardiologists (n=4) | Cardiothoracic surgeons (n=2) |
|---|------------|-----------------------|--------------------------------|-------------------------------|
| <b>Number of children with CHD under their care</b>   |            |                       |                                |                               |
| < 10  | 7 (35%)    | 6 (42.9%)             | 0                              | 1 (50%)                       |
| 10-25   | 3 (15%)    | 3 (21.4%)             | 0                              | 0                             |
| 26-50   | 3 (15%)    | 2 (14.3%)             | 0                              | 1 (50%)                       |
| > 50  | 7 (35%)    | 3 (21.4%)             | 4 (100%)                       | 0                             |
| <b>Importance of neurodevelopmental evaluation and management guidelines for CHD</b>  |            |                       |                                |                               |
| Very important  | 14 (70%)   | 8 (57.1%)             | 4 (100%)                       | 2 (100%)                      |
| Somewhat important  | 4 (20%)    | 4 (28.6%)             | 0                              | 0                             |
| Not important   | 0          | 0                     | 0                              | 0                             |
| Unsure  | 2 (10%)    | 2 (14.3%)             | 0                              | 0                             |
| <b>Awareness of the 2012 AHA guideline on evaluation and management of neurodevelopment in CHD</b>  |            |                       |                                |                               |
| Yes   | 4 (20%)    | 1 (7.1%)              | 2 (50%)                        | 1 (50%)                       |
| No  | 16 (80%)   | 13 (92.9%)            | 2 (50%)                        | 1 (50%)                       |
| <b>DEVELOPMENTAL SURVEILLANCE</b>   |            |                       |                                |                               |
| <b>Identifying children at risk or with developmental problems through observation, identification of parental concerns and taking a comprehensive developmental history.</b> |            |                       |                                |                               |
| <b>Routinely perform developmental surveillance in their clinical practice</b>  |            |                       |                                |                               |
| Yes   | 15 (75%)   | 12 (85.7%)            | 3 (75%)                        | 0                             |
| No  | 5 (25%)    | 2 (14.3%)             | 1 (25%)                        | 2 (100%)                      |
| <b>Healthcare service provider performing the surveillance*</b>   |            |                       |                                |                               |
|   | (n=15)     | (n=12)                | (n=3)                          | (n=0)                         |
| Self  | 15 (100%)  | 12 (100%)             | 3 (100%)                       | 0                             |
| Paediatric neurologist  | 1 (6.7%)   | 1 (8.3%)              | 0                              | 0                             |
| Neurodevelopmental paediatrician  | 4 (26.7%)  | 2 (16.7%)             | 2 (66.7%)                      | 0                             |
| Physiotherapist   | 4 (26.7%)  | 2 (16.7%)             | 2 (66.7%)                      | 0                             |
| Occupational therapist  | 7 (46.7%)  | 6 (50%)               | 1 (33%)                        | 0                             |
| Speech therapist  | 4 (26.7%)  | 3 (25%)               | 1 (33%)                        | 0                             |
| Psychologist  | 0          | 0                     | 0                              | 0                             |
| <b>Risk-stratify children with CHD according to 2012 AHA guideline</b>  |            |                       |                                |                               |
|   | (n= 19)    | (n=13)                | (n=4)                          | (n=2)                         |
| Yes   | 1 (5.3%)   | 1 (7.7%)              | 0                              | 0                             |
| No  | 18 (94.7%) | 12 (92.3%)            | 4 (100%)                       | 2 (100%)                      |
| <b>DEVELOPMENTAL SCREENING</b>  |            |                       |                                |                               |
| <b>The administration of a brief standardised tool to identify child at risk of or presenting with a developmental delay.</b>   |            |                       |                                |                               |
| <b>Children with CHD are routinely referred for developmental screening</b>   |            |                       |                                |                               |
| Yes   | 13 (65%)   | 8 (57.1%)             | 4 (100%)                       | 1 (50%)                       |
| No  | 7 (35%)    | 6 (42.9%)             | 0                              | 1 (50%)                       |
| <b>Ages at which children are referred for developmental screening*</b>   |            |                       |                                |                               |
| 3 months  | 3 (15%)    | 2 (14.3%)             | 1 (25%)                        | 0                             |
| 6 months  | 5 (25%)    | 3 (21.4%)             | 2 (50%)                        | 0                             |
| 9 months  | 2 (10%)    | 2 (14.3%)             | 0                              | 0                             |
| 12 months   | 4 (20%)    | 3 (21.4%)             | 1 (25%)                        | 0                             |
| 18 months   | 4 (20%)    | 3 (21.4%)             | 1 (25%)                        | 0                             |
| 24 months   | 3 (15%)    | 3 (21.4%)             | 0                              | 0                             |
| 30 months   | 3 (15%)    | 3 (21.4%)             | 0                              | 0                             |
| 48 months   | 2 (10%)    | 2 (14.3%)             | 0                              | 0                             |
| All care visits   | 15 (75%)   | 11 (78.6%)            | 3 (75%)                        | 1 (50%)                       |
| <b>Make use of electronic and web-based applications for developmental screening</b>  |            |                       |                                |                               |
| Yes   | 1 (5%)     | 1 (7.1%)              | 0                              | 0                             |
| No  | 16 (80%)   | 11 (78.6%)            | 4 (100%)                       | 1 (50%)                       |
| Feel it is not applicable   | 3 (15%)    | 2 (14.3%)             | 0                              | 1 (50%)                       |

\*Multiple category responses permitted. CHD = Congenital heart disease.

TABLE III: Survey response of practitioners (Part II Developmental evaluation).

| Variable   | All (n=20) | Paediatricians (n=14) | Paediatric cardiologists (n=4) | Cardiothoracic surgeons (n=2) |
|--|------------|-----------------------|--------------------------------|-------------------------------|
| <b>DEVELOPMENTAL EVALUATION</b>  |            |                       |                                |                               |
| <b>Comprehensive process of detailed evaluation and testing aimed at identifying the specific developmental delay or disorder.</b> |            |                       |                                |                               |
| <b>Frequency at which children who are identified as being developmentally at-risk are referred for formal medical evaluation</b>  |            |                       |                                |                               |
| Never  | 0          | 0                     | 0                              | 0                             |
| Rarely (<25%)  | 4 (20%)    | 2 (14.3%)             | 1 (25%)                        | 1 (50%)                       |
| Sometimes (20-25%)   | 6 (30%)    | 5 (35.7%)             | 1 (25%)                        | 0                             |
| Most of the time (50-75%)  | 6 (30%)    | 4 (28.6%)             | 2 (50%)                        | 0                             |
| Almost always (>75%)   | 4 (20%)    | 3 (21.4%)             | 0                              | 1 (50%)                       |
| <b>Referrals to the following healthcare service providers for formal developmental evaluations*</b>                               |            |                       |                                |                               |
| Special medical evaluations (neurologist and neuroimaging)   | 11 (55%)   | 8 (57.1%)             | 2 (50%)                        | 1 (50%)                       |
| Allied health (physio, occupational and speech therapy)  | 15 (75%)   | 9 (64.3%)             | 4 (100%)                       | 2 (100%)                      |
| Interdisciplinary high-risk clinics  | 4 (20%)    | 2 (14.3%)             | 2 (50%)                        | 0                             |
| Perform the evaluation themselves  | 7 (35%)    | 6 (42.9%)             | 1 (25%)                        | 0                             |
| <b>Age period children are referred for formal developmental evaluation*</b>   |            |                       |                                |                               |
| Neonatal period  | 3 (15%)    | 3 (21.4%)             | 0                              | 0                             |
| Infancy  | 14 (70%)   | 10 (71.4%)            | 4 (100%)                       | 0                             |
| Pre-schooler   | 9 (45%)    | 7 (50%)               | 1 (25%)                        | 1 (50%)                       |
| Schooler   | 3 (15%)    | 2 (14.3%)             | 1 (25%)                        | 0                             |
| Adolescence  | 1 (5%)     | 1 (7.1%)              | 0                              | 0                             |
| Transition to adulthood  | 1 (5%)     | 1 (7.1%)              | 0                              | 0                             |
| <b>Circumstances under which children are referred for formal developmental assessment*</b>  |            |                       |                                |                               |
| Parental concerns about development  | 16 (80%)   | 11 (78.6%)            | 3 (75%)                        | 2 (100%)                      |
| Own concerns about development   | 17 (85%)   | 12 (85.7%)            | 4 (100%)                       | 1 (50%)                       |
| Based on developmental screening test results  | 8 (40%)    | 7 (50%)               | 1 (25%)                        | 0                             |
| Children underwent open heart surgery in infancy   | 4 (20%)    | 3 (21.4%)             | 0                              | 1 (50%)                       |
| Children's cyanotic lesions who did not undergo surgery in infancy   | 5 (25%)    | 4 (28.6%)             | 0                              | 1 (50%)                       |
| Presence of a genetic abnormality or syndrome  | 15 (75%)   | 10 (71.4%)            | 3 (75%)                        | 2 (100%)                      |
| CHD with comorbidity**   | 12 (60%)   | 9 (64.3%)             | 2 (50%)                        | 1 (50%)                       |
| <b>Children considered to be developmentally at-risk are referred for developmental evaluation*</b>                                |            |                       |                                |                               |
| Interdisciplinary evaluation   | 9 (45%)    | 9 (64.3%)             | 0                              | 0                             |
| Medical evaluation (genetic testing and neuro-imaging)   | 10 (50%)   | 7 (50%)               | 2 (50%)                        | 1 (50%)                       |
| Allied health services (physio, occupational and speech therapy)   | 11 (55%)   | 6 (42.9%)             | 3 (75%)                        | 2 (100%)                      |
| Other  | 1 (5%)     | 0                     | 1 (25%)                        | 0                             |

\*Multiple category responses permitted. \*\*Comorbidities including prematurity, history of cardiopulmonary resuscitation, need mechanical support, prolonged hospitalisation, and neuro-imaging abnormalities

to identify those children at high risk for developmental delays due to cardiac surgery-related factors such as prolonged cardiopulmonary bypass,<sup>(18,19)</sup> the need for cardiopulmonary resuscitation,<sup>(20)</sup> post-operative seizures,<sup>(21)</sup> and prolonged post-operative hospital length of stay.<sup>(10)</sup>

Risk-stratification serves to identify those children with CHD considered to be at high risk for developmental delay, and

who should be referred directly for formal developmental evaluation.<sup>(2)</sup> Only 5.3% of practitioners risk stratified children using the criteria set out in the 2012 AHA guideline. This was considerably lower than the reported 25% of practitioners risk-stratifying children in the US survey.<sup>(4)</sup> Referral for formal developmental evaluation tended to be based on practitioners' own personal or reported parental concerns about the child's

**TABLE IV: Survey response of practitioners (Part III Developmental evaluation and access to intervention therapies).**

| Variable   | All (n=20) | Paediatricians (n=14) | Paediatric cardiologists (n=4) | Cardiothoracic surgeons (n=2) |
|--|------------|-----------------------|--------------------------------|-------------------------------|
| <b>DEVELOPMENTAL EVALUATION</b>  |            |                       |                                |                               |
| <b>Detailed evaluation and testing using a comprehensive, standardised developmental assessment tool aimed at making a developmental diagnosis and planning care.</b>          |            |                       |                                |                               |
| <b>Are the required medical specialists available to complete formal developmental evaluations?</b>  |            |                       |                                |                               |
| Yes  | 12 (60%)   | 8 (57.1%)             | 2 (50%)                        | 2 (100%)                      |
| No   | 4 (20%)    | 2 (14.3%)             | 2 (50%)                        | 0                             |
| Do not know  | 4 (20%)    | 4 (28.6%)             | 0                              | 0                             |
| <b>Reasons for not referring children for formal developmental evaluation*</b>   |            |                       |                                |                               |
| Family resistance  | 0          | 0                     | 0                              | 0                             |
| Unaware of recommended referral guidelines   | 5 (25%)    | 4 (28.6%)             | 0                              | 1 (50%)                       |
| Medical specialists required are not available   | 8 (40%)    | 5 (35.7%)             | 3 (75%)                        | 0                             |
| Children already accessing developmental care  | 3 (15%)    | 1 (7.1%)              | 0                              | 2 (100%)                      |
| Children already accessing intervention therapies  | 6 (30%)    | 5 (35.7%)             | 0                              | 1 (50%)                       |
| Financial concerns of the family   | 4 (20%)    | 4 (28.6%)             | 0                              | 0                             |
| Travelling distances for families to access services   | 8 (40%)    | 6 (42.9%)             | 2 (50%)                        | 0                             |
| Other  | 2 (10%)    | 1 (7.1%)              | 1 (25%)                        | 0                             |
| <b>Are parents and primary care providers informed of the need for ongoing screening for children identified to be at risk of developmental difficulties?*</b>                 |            |                       |                                |                               |
| Yes  | 16 (80%)   | 10 (71.4%)            | 4 (100%)                       | 2 (100%)                      |
| No   | 4 (20%)    | 4 (28.6%)             | 0                              | 0                             |
| <b>ACCESS TO INTERVENTION THERAPIES</b>  |            |                       |                                |                               |
| <b>Early intervention therapies include physiotherapy, occupational therapy and speech therapy services. Children may additionally require the services of a psychologist.</b> |            |                       |                                |                               |
| <b>Intervention therapies that children at risk or presenting with developmental delays are referred to*</b>   |            |                       |                                |                               |
| Physiotherapist  | 15 (75%)   | 9 (64.3%)             | 4 (100%)                       | 2 (100%)                      |
| Occupational therapist   | 20 (100%)  | 14 (100%)             | 4 (100%)                       | 2 (100%)                      |
| Speech therapist   | 14 (70%)   | 9 (64.3%)             | 3 (75%)                        | 2 (100%)                      |
| Psychologist   | 3 (15%)    | 3 (21.4%)             | 0                              | 0                             |
| <b>Distance, on average, parents with children with CHD must travel to access intervention therapies and child support services</b>  |            |                       |                                |                               |
| Less than 50 km  | 4 (23.5%)  | 4 (36.4%)             | 0                              | 0                             |
| Greater than 50 km   | 13 (76.5%) | 7 (63.6%)             | 4 (100%)                       | 2 (100%)                      |
| <b>Perceived barriers limiting access of children and their families to intervention therapies*</b>  |            |                       |                                |                               |
| Availability of therapy services   | 15 (75%)   | 11 (78.6%)            | 4 (100%)                       | 0                             |
| Traveling distance to therapy services   | 15 (75%)   | 9 (64.3%)             | 4 (100%)                       | 2 (100%)                      |
| Parents feel it is unnecessary, their child is doing well  | 7 (35%)    | 6 (42.9%)             | 1 (25%)                        | 0                             |
| Cost of early intervention therapies   | 5 (25%)    | 4 (28.6%)             | 1 (25%)                        | 0                             |
| Time required from parents to take children to therapy   | 8 (40%)    | 5 (35.7%)             | 2 (50%)                        | 1 (50%)                       |
| Time required to adhere to home exercise programmes  | 8 (40%)    | 6 (42.9%)             | 1 (25%)                        | 1 (50%)                       |
| Loss of income when taking child therapies   | 8 (40%)    | 7 (50%)               | 1 (25%)                        | 0                             |
| Other (siblings at home)   | 2 (10%)    | 1 (7.1%)              | 1 (25%)                        | 0                             |

\*Multiple category responses permitted.

development. Risks specifically related to CHD itself, such as open-heart surgery in infancy and cyanotic lesions, only accounted for around a quarter of the referrals for formal developmental evaluation. Referral practices tended to be reactive, based on the presence of developmental problems or

noted concerns, rather than proactively based on specific CHD-related risks. This would suggest that practitioners were unaware of the specific CHD-related developmental risks and the criteria for referral for formal developmental evaluation.<sup>(4)</sup> The lack of risk stratification practices in the current study



would mean that a considerable number of children at high risk for developmental delay were not being referred early for formal developmental evaluation. Late referral for formal developmental evaluation would, in turn, also result in the delayed referral to intervention therapies. These findings are consistent with those of the US survey.<sup>(4)</sup>

The 2012 AHA guideline recommends that children with CHD undergo periodic developmental screening with a brief standardised screening tool at 9, 18, 30 and 48 months.<sup>(2,11)</sup> Only 65% of practitioners referred children for developmental screening. Screening practices were extremely variable, with referral occurring most often ( $\leq 25\%$ ) at 6, 12 and 18 months of age. Very few children ( $\leq 15\%$ ) were referred for developmental screening at or after the age of 2 years. These screening practices are likely to result in children with mild developmental delays, behavioural problems and inattention being missed.<sup>(1,10)</sup> Contrastingly, many practitioners ( $n=15$ ; 75%) reported referring children for developmental screening at every care visit. This screening practice would unnecessarily increase the workload on already over-extended and understaffed healthcare services, and would furthermore limit the number of children who could be accommodated for developmental screening.<sup>(17,22)</sup> Referral for developmental screening in the current study was far lower than the 90% of practitioners in the US survey who reported regularly referring children for developmental screening. US practitioners also referred more children aged two years and older for developmental screening.<sup>(4)</sup>

A single practitioner reported using electronic web-based developmental screening applications. Neurodevelopmental experts are of the opinion that it has become imperative to consider the use of innovative technologies to facilitate remote developmental screening and therapy interventions in the CHD population – to counteract the high loss to developmental follow-up.<sup>(12,23-26)</sup> Electronic developmental screening questionnaires could be completed by parents at home and sent on to the practitioner to facilitate remote screening of the child's developmental status.<sup>(27)</sup>

Less than a quarter of practitioners ( $n=4$ ; 20%) referred at-risk children for formal developmental evaluation, which would include the administration of a comprehensive, standardised developmental test.<sup>(2)</sup> Children were referred to allied health services ( $n=15$ ; 75%) and to medical specialists such as a paediatric neurologists or neurodevelopmental paediatricians ( $n=11$ ; 55%) for formal developmental evaluation. Based on local experience it would be unlikely that a comprehensive,

standardised developmental test would be administered at formal developmental evaluations, due to time constraints, a lack of testing expertise and the unavailability of these expensive imported developmental tests. Children were rarely referred for developmental evaluation as scholars or adolescents. This is of concern, as one in three children with CHD have educational concerns,<sup>(7,21)</sup> and up to 50% of school-aged children are reported to require intervention therapies and remedial educational services.<sup>(6,7,28)</sup>

Around a third of practitioners ( $n=6$ ) did not refer children for formal developmental evaluation if they were already accessing intervention therapies. The US survey had a similar finding.<sup>(4)</sup> Intervention therapies are important in effectively addressing developmental delays, but do not replace the need for concurrent formal developmental evaluation.<sup>(4)</sup> Barriers to formal developmental evaluation are consistent with previous reports, and include a lack of access to the necessary medical specialists, including paediatric neurologists and neurodevelopmental paediatricians ( $n=8$ ; 40%), and the distance children and their families have to travel to access services ( $n=8$ ; 40%).<sup>(4,14,29)</sup>

Only 20% of practitioners referred children to an interdisciplinary clinic for developmental evaluation, despite an interdisciplinary approach being strongly advocated.<sup>(13,14)</sup> The prohibitive infrastructure and human resource costs of establishing dedicated cardiac neurodevelopmental clinics have resulted in the cardiac care community in both the US and Canada recommending that existing, well established interdisciplinary high-risk clinics be used for cardiac neurodevelopmental follow-up.<sup>(6,12-14)</sup> The lack of utilisation of existing high-risk interdisciplinary clinics in the current study is of concern, as this is likely to be the only feasible option for providing cardiac neurodevelopmental follow-up in South Africa.

Children presenting with developmental delays were referred to occupational therapy (100%), physiotherapy (75%) and speech therapy (70%). Children were mostly screened before the age of two years for developmental delays, where motor delays and language deficits are known to be more common.<sup>(1)</sup> Screening practices would have suggested greater numbers of referrals to physiotherapy and speech therapy to address these developmental delays. It must also be considered that the referral pattern may reflect the therapy services available to children in central South Africa.<sup>(30)</sup> Children with CHD are at increased risk of behavioural and social difficulties, and are 3 - 4 times more likely to present with inattention and hyperactivity than the general population.<sup>(2,7,19,21)</sup> Only 15% of participants referred children to a psychologist. Reasons for the low number

of referrals to psychologists may be due to the small number of school-aged and adolescent children screened and evaluated for developmental and behavioural difficulties. Furthermore, psychologists are a very limited resource in the healthcare sector globally.<sup>(31,32)</sup>

Perceived barriers to accessing intervention therapies included the distance families had to travel to access therapy, the time required from parents to take their child for therapy, the time required to adhere to the prescribed home exercise programmes, and the loss of income suffered by parents who had to take their children to therapy appointments. Most parents (76.5%) stayed more than 50 kilometres from their closest cardiac centre, where intervention therapies were offered. The noted barriers are consistent with those in the scant published literature on the feasibility and practicability of providing developmental interventions to children with CHD.<sup>(25,26)</sup>

### Limitations

The study findings need to be interpreted in the light of several limitations. Though the survey response rate at 46.7% is higher than the reported average response rate of 33% for online surveys, the sample size was small.<sup>(33)</sup> Reasons for the non-responses may have included incorrect email contact addresses, emails being caught up in spam filters, practitioners not considering the survey to be of interest or of relevance to them, and practitioners not having the time to complete the survey due to their already overburdened work schedules.<sup>(34,35)</sup>

The findings are specific to the population of practitioners in central South Africa and may not be generalisable to practitioners caring for children with CHD in South Africa or globally. The small sample size limited the ability to compare the responses for the various practitioner categories. The online survey was only available for 6 weeks, which may have limited the number of responses received.

### Recommendations

A national survey in a larger sample of cardiac care practitioners is needed to better determine practitioners' awareness of the 2012 AHA guidelines, as well as their neurodevelopmental evaluation and referral practices. The feasibility of remote web-based developmental screening applications accessible via a smartphone, requires further investigation.

### CONCLUSION

Most practitioners in central South Africa are unaware of the 2012 AHA guideline and the developmental risks faced by children with CHD. Current evaluation practices are sub-

optimal and are likely to result in children with CHD at high risk for developmental delays not being identified and referred for early formal developmental evaluation and intervention therapies. Strategies to raise awareness of the 2012 AHA guideline among cardiac care practitioners need to be devised.

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### DISCLAIMER

The views expressed in this article are those of the authors alone, and are not a reflection of the official position of the institution represented.

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