

Quality of life can be good after slide tracheoplasty for long-segment tracheal stenosis

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Key question: How good is the health-related quality of life (HRQoL) of children with repaired long-segment tracheal stenosis (LSTS)?

Key findings: Mean scores did not differ from those of healthy norms

Non-cardiac co-morbidities were associated with lower scores

Take home message: Children with repaired LSTS can have excellent HRQoL but non-cardiac co-morbidities are a risk factor for poorer outcome

There is no central image for this article.

Abstract

Objectives

To measure 'health-related quality of life' (HRQoL) in children following slide tracheoplasty for long-segment tracheal stenosis (LSTS) and to explore the relationship of co-morbidities and parental mental health with HRQoL outcomes.

Methods

A cross sectional study was undertaken with children who had undergone slide tracheoplasty. Participants included parents and children (age 5-15 years) recruited over a 13-month period, who were asked to complete validated measures of HRQoL, development and behavior. Scores were compared to published norms.

Results

Forty-two children (male 69%; n=29) were included; mean age 5.3 (S.D. 3.5) years, and a mean follow up of 45 (range 4-179) months. Mean total HRQoL scores for children with repaired LSTS did not differ from those of healthy norms other than for children aged 13-23 months but 10 children (24%) had scores >2 SD below the mean for healthy children. HRQoL was poorer in children with non-cardiac congenital co-morbidities than in those with isolated LSTS (mean scores = 60.34 +/-17.19 and 85.52 +/-12.19, respectively, p=0.01). There was good agreement between children's and parents' scores, although children rated their HRQoL as better than their parents did. Anxious parents rated their children's HRQoL as significantly worse than non-anxious parents (p<.001).

Conclusions

Older children with isolated LSTS can have excellent HRQoL after surgery. Younger children, at an earlier time point post-operatively, and those with non-cardiac congenital co-morbidities have poorer HRQoL. Further longitudinal evaluation is required to identify

psychosocial (including parental) predictors of outcome which may inform, or be amenable to, intervention.

Key words

Long-segment tracheal stenosis, quality of life, development, children

Level of evidence

Level IV

Acknowledgements:

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Introduction

Long-segment tracheal stenosis (LSTS) is a rare, serious and potentially life-threatening condition characterised by severe narrowing of the trachea spanning more than two-thirds of its overall length.[1] The true incidence of LSTS is unknown [2] but the cause can be congenital or acquired. [3] There is a wide spectrum of morphology, including the site and length of stenosis, bronchial involvement and the presence of complete tracheal cartilage rings. [4] The condition is commonly diagnosed during investigation of other co-morbidities, especially cardiac anomalies which may be present in up to 70% of patients. [5] The surgical repair of choice today is slide tracheoplasty, first performed in the 1980s, which has lower rates of associated mortality and morbidity compared with previously used techniques such as pericardial patch tracheoplasty, autograft and homograft insertion and stenting. [6]

Published data on LSTS focus on surgical and medical management and clinical outcomes, with as yet little attention given to health-related quality of life (HRQoL) or other developmental and psychological parameters. Despite the significant risk factors for morbidity and mortality following repair of LSTS, operative survival rates continue to improve and internationally range from 67% to 95% [3, 5, 7, 8] but reducing mortality rates mean that there are, paradoxically, likely to be increasing numbers of children living with some degree of morbidity following repair of LSTS. It is well documented that children with chronic health conditions are at increased risk for impaired HRQoL and poorer psychological adjustment [9] and evaluating these parameters is now indicated in children with repaired LSTS.

The aim of this study was to measure HRQoL and development in a cohort of children who had undergone repair of LSTS with slide tracheoplasty and to compare them with published norms for healthy children. We further explored the relationship of co-morbidities and parental mental health with HRQoL outcomes.

Methods

Ethical approval and consent processes

The study was approved by the National Research Ethics Committee (11/LO/0132). Parents provided written consent for their own participation and that of their child and children able to give assent for their participation did so.

Participants

All children who had undergone a slide tracheoplasty for LSTS and were under follow-up at our institution and their parents were eligible for participation. Parents and older children were considered eligible if comprehension of English language was sufficient for completion of the questionnaires, with assistance if necessary.

Procedure

Eligible children and their families were identified from a prospectively recorded database of children with LSTS and invited to participate in the study by the tracheal clinical nurse specialist or a medical student over a 13-month period. Participants either completed questionnaires when attending for clinical review or through postal invites and returns.

Measures

Quality of life

Perceived HRQoL was assessed with the PedsQL4.0 [10, 11], a generic measure existing in parent-proxy form for ages 1-12 months, 13-23 months, 2-4 years, 5-7 years, 8-12 years and 13-18 years and in child/teen completed form for the three older age groups. The two infant versions comprise 36 and 45 questions respectively, each having five sub-scales: physical functioning, physical symptoms, emotional functioning, social functioning and cognitive functioning. Questionnaires for children aged 2-18 years comprise 23 items and four subscales: physical, emotional, social and nursery/school functioning. For all age groups a

total score, physical health summary score and psychosocial summary score can be calculated, each with a range of 0-100 where higher scores equate to better perceived HRQoL.

Development and behaviour

For children under five years of age, development and behaviour were assessed with the Ages and Stages Questionnaire (ASQ-3) [12] and the Ages and Stages Social-Emotional Questionnaire (ASQ-SE). [13] The ASQ-3 is a parent-completed developmental screening instrument and exists as 21 questionnaires covering the age-range of 2-60 months. Each questionnaire consists of 30 items focused on five areas of development: communication, gross motor, fine motor, problem solving and personal-social. For each item parents are asked whether children have achieved the particular milestone. Total scores are computed for each domain and compared to a cut-off score, which varies according to the age of the child and domain. Scores below the cut-off equate to scores more than two standard deviations below the mean and may indicate developmental delay. The ASQ-SE is a parent-completed screening questionnaire specifically focusing on social and emotional behaviour. It exists as eight different questionnaires covering the age-range of 6–60 months and the number of items on the questionnaires ranges from 19-33. A total score is computed by summing individual item scores and children scoring above the cut-off (which varies depending on the age of the child) are considered to have the potential for social-emotional concerns.

Behaviour of children aged 5 years and older was assessed with the Strengths and Difficulties Questionnaire (SDQ) [14], a 25-item questionnaire completed by parents, with a self-report version for children of 11 years and older. There are five subscales, each with five items: emotional symptoms, conduct problems, peer relationships, hyperactivity/inattention and prosocial behaviour. Scores for each subscale range from 0-10 and a total difficulties score is computed by summing scores from all scales except for the

prosocial scale. Total scores of 17-19 are considered to be high and scores of 20-40 are very high. The SDQ was introduced later in the study and was therefore only administered to a subset of eligible children.

Parental mental health and family functioning

Parental mental health was assessed with the Hospital Anxiety and Depression Scale (HAD) [15], a 14-item questionnaire with two subscales measuring anxiety (seven items) and depression (seven items). Total scores for each subscale range from 0-21, with scores of 8–10 indicating borderline levels of anxiety or depression and scores of 11 or more indicating clinical levels.

Family functioning was assessed with the 12-item General Functioning subscale (GF12) of the McMaster Family Assessment Device. [16] The GF12 has been validated as a single index measure to assess family functioning and comprises six positively worded items and six negatively worded items to capture both healthy and unhealthy family functioning. Each item is scored on a 4-point scale and negative items are reverse scored. The summed score is divided by the number of items to give a total score ranging from 1.0 (best functioning) to 4.0 (worst functioning), with a score of 2.0 or more indicating concern about family functioning.

All questionnaires have acceptable reliability and validity and have been widely used in studies of healthy children and children with a range of chronic and acute health conditions.

Statistical analysis

Descriptive statistics were used to explore the study sample. One sample t-tests were used to compare HRQoL scores of children who had undergone repair of LTST in each age group with scores for healthy norms. [10, 11, 17] Chi-square (or Fisher's exact test if assumptions for the Chi-square test were not met), Mann-Whitney and Kruskal-Wallis tests were used to

compare PedsQL, ASQ, SDQ, GF-12 and HADs scores between those with and without congenital co-morbidities. As this was an exploratory analysis no correction for multiple comparisons was made and a p value of <0.05 was considered statistically significant. Spearman bivariate correlations were undertaken to identify significant associations between medical, demographic and parental variables and HRQoL, development and behaviour scores. Due to small sample sizes within individual age bands, PedsQL scores from all age groups were combined for analyses other than the comparison with healthy norms. Statistical analysis was performed using SPSS version 22.0 (SPSS, Inc., Chicago, Illinois). For descriptive purposes, we have defined younger children as those younger than 5 years of age and older children as those of 5 years and older.

Results

Fifty-seven children met the inclusion criteria, all of whom had undergone slide tracheoplasty for LSTS between 1998 and 2012, and parents of 54 agreed to participate. The surgical approach in all children was via a median sternotomy on cardiopulmonary bypass as previously described [5]. No information was provided by those who did not agree to take part. Questionnaires were returned by families of 42 children (74% of those eligible to participate). Demographic and medical data are provided in Table 1. Seven of the 11 children (64%) under 2 years of age had a congenital co-morbidity +/- a cardiovascular anomaly compared with 10 of the 31 children (32%) aged 2 years and older. All children with other co-morbidities had undergone other surgery. The 15 children who did not participate did not differ significantly from those who did participate on any demographic or medical variables.

Quality of life

Parents of 41 children completed the PedsQL (one child had too much missing data to be included) and 19 of the 21 (90%) eligible children completed the self-report form. Scores for children in each age group are shown in Table 2, together with published norms for healthy

children. [10, 11, 17] Scores did not differ from healthy norms other than for the children aged 13-23 months where scores on all scales other than the social and cognitive scales were significantly ($p < 0.05$) lower. Ten of the 41 children (24%) had total HRQoL scores of more than 2 SD below the mean for healthy children, seven of whom had congenital (non-cardiac) co-morbidities and two had a cardiovascular anomaly together with a non-cardiac co-morbidity. There was good agreement between children and parents on all domains of HRQoL. Children consistently rated their HRQoL as better than their parents did but the differences were not significant.

Small sample sizes precluded statistical analysis of the subgroups. Overall, however, there were no differences on any PedsQL scores for those children who had no congenital comorbidities ($n=6$) and those who had any congenital abnormality ($n=35$). However, as shown in Figure 1, looking at children with a congenital co-morbidity, those with a cardiovascular anomaly only ($n=18$) had better HRQoL in all domains than those with a non-cardiac congenital co-morbidity only ($n=8$). For the total group there were no significant correlations between any demographic or clinical variables and HRQoL scores.

Development and behaviour

ASQ-3 questionnaires were completed for 15 of the 21 children (71%) aged less than 5 years. Numbers of children scoring below the cut-off for each subscale are shown in Table 3. Nine (60%) children scored below the cut-off on at least one subscale, all of whom had either a cardiovascular anomaly ($n=1$), a non-cardiac congenital comorbidity ($n=5$) or both ($n=3$).

ASQ-SE questionnaires were completed for 17 of the 20 eligible children (85%), 2 of whom (12%) scored above the cut-off.

Parents of nine of the 22 eligible children completed the SDQ (due to its later introduction) and all four children aged 11 years and older completed the self-report version. Median subscale and total scores are shown in Table 3. All scores for both parent and self-report measures were within the normal range with the exception of two children (22%) who had hyperactivity scores in the borderline range on parent report.

Parental mental health and family functioning

Parents of 34 children completed the HAD, of whom six (18%) and three (9%) had borderline levels of anxiety and depression respectively and 11 (32%) and two (6%) had clinical levels of anxiety and depression respectively, with both parents who had clinical levels of depression also having clinical levels of anxiety. The proportion of parents with clinical levels of anxiety did not differ between those parents whose children had a congenital comorbidity and those whose children did not. The median score on the GF-12 was 1.5 (IQR: 1.17-1.75), with two (5%) of the 39 parents who completed the measure scoring 2 or more.

Parents who had clinical levels of anxiety rated their child's HRQoL as lower than those parents who did not have clinical levels of anxiety (Figure 2).

Discussion

Long-segment tracheal stenosis is a rare condition which is increasingly being repaired successfully. To date the focus has been on mortality and morbidity, and as far as we are aware there are no previous reports of HRQoL or development following repair with slide tracheoplasty. Our results are encouraging and indicate that older children in particular – and therefore those who are further from their slide tracheoplasty – can have excellent HRQoL which does not differ from their healthy peers, particularly if they have isolated tracheal stenosis rather than tracheal stenosis in conjunction with non-cardiac congenital co-

morbidities. Furthermore, older children had low rates of behavior problems on both self and parent rating, with borderline problems only being reported on the hyperactivity scale by parents of two children. However, findings for younger children and those with co-morbidities suggest that some children have poor HRQoL, evidenced by the fact that a quarter of the children had total HRQoL scores which were more than 2 SD below the mean for the healthy population. Moreover, all of the school-aged children with a non-cardiac congenital co-morbidity were receiving special education and had the lowest HRQoL scores in the school-aged population.

In general, HRQoL scores for the younger children were lower than healthy norms, particularly for those children aged 13-23 months. Items on the PedsQL for the youngest two age groups are aligned to development and the poor results are supported by the results on the ASQ-3, on which more than half of the younger children had scores below the cut-off in at least one area of development. Gross motor skills were the domain with the greatest impairment, with half of the children scoring poorly on this scale. This latter finding is not surprising given the recognised impact of invasive surgery and hospitalisation on children's psychomotor development [18] - some of the younger children were less than a year from the time of their surgery. Although age was not significantly correlated with outcome, it is also noteworthy that a higher proportion of children under 2 years of age had a congenital co-morbidity +/- a cardiovascular anomaly. Some of these co-morbidities are known to be associated with developmental delay, such as Down syndrome [19] and CHARGE syndrome, [20] but others have no known association with developmental impairment. Longitudinal follow-up of these children will be important to determine whether they catch-up with their healthy peers over time. In contrast to some previous findings suggesting that children with congenital heart disease have poorer HRQoL than their healthy peers, [21] children with LSTS and a cardiovascular anomaly as the only co-morbidity did not have poorer HRQoL than healthy norms. This is likely because published studies focus on more complex cardiac lesions, which are not typically seen in children with LSTS, where the

cardiovascular anomaly is typically a left pulmonary artery sling, considered a relatively 'minor' anomaly.

In common with other studies of HRQoL in children with chronic health conditions, [22] there was good agreement between parents and their children on all domains of HRQoL other than for social HRQoL. Previous research has identified better agreement on more visible or objective domains of HRQoL[22] and it is not surprising that parents may be less aware of how their child functions socially compared with aspects such as physical functioning. Children rated their HRQoL as better than their parents did, supporting findings for other groups of children [23, 24] including those with laryngotracheal stenosis, [25] although differences were not significant. Such findings do, however, support the view that both children and parents should be asked about a child's HRQoL. Parental mental health is also an important consideration - our findings indicated that parents with clinical levels of anxiety rated their child's HRQoL as poorer than parents who did not have clinical levels of anxiety. Similar findings concerning the role of parental mental health in reporting of their child's HRQoL have been identified for other groups of children [26] and indicate the importance of measuring parental mental health when evaluating HRQoL, particularly if children are unable to provide information themselves about their HRQoL. Interestingly, however, family functioning scores indicated good overall functioning and a low proportion of scores suggesting family dysfunction. Other studies have reported higher rates of family dysfunction where children have a chronic condition [27] and one explanation for the comparatively low rates in the current study is that in most cases the child's condition was stable and relatively 'low maintenance' with all surgery having taken place, often several years previously.

There are a number of limitations which need to be taken into consideration. Firstly, LSTS is a rare condition and the sample size was therefore small, precluding further analysis, although HRQoL data were collected for almost three-quarters of eligible patients. Whilst

the sample was representative in terms of demographic and clinical data of the whole LSTS population at our institution, it was heterogenous with respect to time since surgery and the presence of cardiac and non-cardiac comorbidities. We were also not able to look at the potential impact of other factors on outcome, such as whether or not the child and family were seen by a psychologist, which would be important to address in future longitudinal evaluation. There may also be a bias in terms of those families who agreed to participate but who did not return questionnaires – these were all children who were not seen in the clinic during the 13 months of data collection, which may have indicated that they were healthier and not requiring such frequent follow-up compared with those who were seen. Data were collected at a single site and whilst our institution provides the national specialist service for the treatment of LSTS in the UK, our findings may have limited generalisability beyond the UK. Furthermore, the cross-sectional design limited our ability to determine causality. The measure of behavior for children aged 5 years and older was introduced part-way through the study and so data are only available for a proportion of the eligible children. Finally, HRQoL was assessed with a generic HRQoL measure. The gold standard is to use both a generic (enabling comparison with other groups of children, including healthy norms) and a disease-specific (to be sufficiently sensitive to enable disease-specific characteristics to be detected) measure [28] but as yet there is no disease-specific measure available for children with conditions affecting the trachea. It is therefore likely that some of the more nuanced ways in which HRQoL may be affected by LSTS were not detected.

Conclusion

Routine assessment of HRQoL is increasingly used to monitor outcomes following complex interventions and evaluate treatment effectiveness. Our findings offer encouragement for children undergoing slide tracheoplasty for LSTS but also highlight the increased risk for poorer outcomes in children with additional non-cardiac co-morbidities. Longitudinal follow-up is now required to identify physical and psychosocial predictors of poor outcome so that interventions can be targeted for those at greater risk. Similarly, identifying psychosocial

factors associated with better outcomes can also inform interventions. Follow-up should include parental assessment and both child and parent reporting where feasible as part of the holistic care of children and families.

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Conflict of interest statement

None of the authors have any conflict of interest to declare.

Figure legends

Figure 1

Figure 2

Table 1: Demographic and clinical data

Variable	Number/Mean/ Median/Range/%
Gender: Number of males	29 (69%)
Age: mean (SD) (years)	5.3 (3.5)
Age range: (years)	0.5 -15.1
Ethnicity: White	34 (81%)
Asian	2 (5%)
Black	5 (12%)
Mixed	1 (2%)
Education:	
Too young for formal education	19(45%)
Receiving education	23 (55%)
Main stream school – no additional support	16 (70%)
Special educational needs	7 (30%)
Co-morbidities:	
No co-morbidities	7 (17%)
Cardiovascular anomaly	18 (43%)
Other congenital co-morbidity	8 (19%)
Cardiovascular anomaly + other congenital co-morbidity	9 (21%)
Cardiovascular anomaly	27 (64%)
Tetralogy of Fallot	5 (19%)
Ventricular septal defect	2 (7%)

Left pulmonary artery sling	20 (74%)
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Congenital co-morbidities other than cardiovascular anomaly	17 (40%)
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Agenesis of right lung	2 (12%)
Anorectal malformation	4 (24%)
VACTERL	2 (12%)
Down syndrome	3 (18%)
CHARGE syndrome	1 (6%)
Extreme prematurity +/- other co-morbidities	2 (12%)
Other	3 (18%)
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Age at slide tracheoplasty : median (range) (months)	6 (0-71)
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Time since tracheoplasty: median (range) (months)	45 (4-179)
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Post-operative complications ^a	22 (52%)
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Phrenic nerve palsy	3 (14%)
Mediastinitis	5 (23%)
Stenosis	4 (18%)
Pneumothoraces	6 (27%)
Chylothorax	2 (9%)
Respiratory problems	3 (14%)
Cardiac arrest	1 (5%)
Vocal chord palsy	2 (9%)
Acute renal failure	1 (5%)
Post-pericardiotomy syndrome	1 (5%)

^aComplications are not mutually exclusive

Table 2: Child and parent-reported PedsQL subscales and total scores (SD) for children who had undergone repair of long-segment tracheal stenosis (LSTS) and healthy norms and correlations between child and parent ratings

	LSTS patients	Healthy norms ^z
Infant (0-12 months)	(n=3)	(n=246)
Physical functioning	86.11 (12.03)	87.54 (11.16)
Physical symptoms	76.67 (7.64)	83.45 (10.39)
Emotional	66.66 (9.56)	76.59 (13.71)
Social	87.50 (16.53)	89.62 (14.87)
Cognitive	79.17 (21.95)	83.11 (20.65)
Physical summary	80.21 (9.15)	84.98 (9.45)
Psychosocial summary	73.75 (10.90)	80.47 (12.64)
Total score	76.39 (9.03)	82.47 (9.95)
Infant (13-23 months)	(n=8)	(n=141)
Physical functioning	65.76 (26.76)	90.32 (8.96)
Physical symptoms	71.25 (14.88)	87.54 (9.29)
Emotional	67.61 (8.13)	78.60 (12.80)
Social	81.09 (20.24)	91.14 (10.77)
Cognitive	61.85 (34.75)	84.65 (15.76)
Physical summary	68.53 (21.24)	88.84 (7.68)
Psychosocial summary	68.99 (13.83)	83.12 (11.02)
Total score	68.51 (14.78)	85.55 (8.74)
Toddler (2-4 years)	(n=9) ^x	(n=2900)
Physical functioning	78.82 (26.14)	89.82 (15.43)
Emotional	83.33 (13.69)	84.26 (14.24)

Social	83.33 (21.65)	88.54 (15.58)
Nursery	78.52 (22.45)	-
Psychosocial summary	81.88 (17.99)	86.56 (12.31)
Total score	81.20 (20.03)	87.86 (12.19)
Parent of children 5-18 years	(n=21)	(n=8713)
Physical functioning ^a	84.82 (20.59)	89.06 (12.27)
Emotional ^b	80.48 (21.73)	78.28 (15.54)
Social ^c	86.90 (16.54)	86.82 (15.42)
School ^d	79.52 (22.47)	81.52 (16.09)
Psychosocial summary ^e	82.14 (17.58)	82.21 (12.67)
Total score ^f	83.38 (16.88)	84.61 (11.19)
Children 5-18 years	(n=19)	(n=5079)
Physical functioning ^a	90.13 (9.95)	88.51 (11.62)
Emotional ^b	83.68 (18.55)	78.49 (19.94)
Social ^c	87.37 (15.22)	87.65 (16.46)
School ^d	83.16 (16.77)	78.87 (15.89)
Psychosocial summary ^e	84.69 (14.16)	81.84 (13.21)
Total score ^f	86.59 (12.42)	83.89 (11.84)

^zSources for healthy norm data [10, 11, 17]

^xOne child in this age group was excluded due to missing data

Correlations between child and parent scores

a: $r=.676$; $p=.01$ b: $r=.799$; $p<.001$ c: $r=.420$; $p=.074$

d: $r=.750$; $p<.001$ e: $r=.789$; $p<.001$ f: $r=.856$; $p<.001$

Table 3: ASQ-3 and SDQ scores for children who had undergone repair of long-segment tracheal stenosis and healthy norms

ASQ-3: Numbers of children scoring below the cut-off on each subscale		
Subscale (n=15)	Number scoring below cut-off (%)	Healthy norm values – proportion scoring below cut-off (%)^b
Communication	6 (40)	16
Gross motor	7 (47)	16
Fine motor	5 (33)	16
Problem solving	4 (27)	16
Personal-social	4 (27)	16
Scoring below cut-off in at least one domain	9 (60)	-

SDQ: Mean subscale and total scores		
Subscale	Median (25th/75th centile)	Mean healthy norm values^b (SD)
Parent-reported (n=9)		n=10,298
Emotional	1.0 (0-2.5)	1.9 (2.0)
Conduct	1.0 (0-1.0)	1.6 (1.7)
Hyperactivity	4.0 (1.5-5.5)	3.5 (2.6)
Peer relationships	1.0 (0-1.5)	1.5 (1.7)
Prosocial	910.0 (8.5-10.0)	8.6 (1.6) ^a
Total score	6.0 (3.0-9.5)	8.4 (5.8)

Child-reported (n=4)		n=4228
Emotional	0.5 (0-1.75)	1.4 (1.9)
Conduct	0.5 (0-1.0)	0.9 (1.6)
Hyperactivity	3.50 (0.75-4.75)	2.9 (2.8)
Peer relationships	3.0 (0.5-4.0)	1.4 (1.8)
Prosocial	10.0 (7.75-10.0)	7.2 (2.4) ^a
Total score	7.5 (1.25-11.5)	6.6 (6.0)

^aHigher scores indicate better functioning on the prosocial scale

^bPublished data have been used to provide healthy norm values for the ASQ-3 [29] and SDQ [30].

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