ORIGINAL ARTICLE

# Short term and long term health related quality of life after congenital anorectal malformations and congenital diaphragmatic hernia

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Tables A and B and Figures A and B are available on the *ADC* website (www.archdischild.com/supplemental)

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**Aims:** To examine short term and long term health related quality of life (HRQoL) of survivors of congenital anorectal malformations (ARM) and congenital diaphragmatic hernia (CDH), and to compare these patients' HRQoL with that of the general population.

**Methods:** HRQoL was measured in 286 ARM patients and 111 CDH patients. All patients were administered a symptom checklist and a generic HRQoL measure. For the youngest children (aged 1–4) the TAIQOL (a preliminary version of the TAPQOL) was used, for the other children (aged 5–15) the TACQOL questionnaire, and for adults (aged >16) the SF-36.

**Results:** As appeared from the symptom checklists, many patients remained symptomatic into adulthood. In the youngest ARM patients (aged 1–4 years), generic HRQoL was severely affected, but the older ARM patients showed better HRQoL. In the CDH patients, the influence of symptoms on HRQoL seemed less profound. The instruments we used revealed little difference between adults treated for ARM or CDH and the general population.

**Conclusions:** These results show that for two neonatal surgical procedures, improved survival does not come at the expense of poor HRQoL in adults. Even though there is considerable suffering in terms of both morbidity and mortality in the youngest group, the ultimate prognosis of survivors of the two studied congenital malformations is favourable. This finding can be used to reassure parents of patients in need of neonatal surgery for one of these conditions about the prospects for their child.

Improved survival among neonates with previously life threatening congenital anomalies has created new dilemmas in neonatal surgery. Mere survival is no longer sufficient; the child's future health related quality of life (HRQoL) is equally important. Moreover, the large amount of economic resources currently spent on neonatal care, coupled with increasing budget constraints, has evoked the question of whether the effects of a given treatment are worth the costs.[1–3] There is growing political interest in evidence based, cost effective medicine, including paediatric surgery.

Recently, we reviewed the literature dated 1989–98 on the prognosis in terms of HRQoL of newborns operated on for the congenital anomalies listed by Ravitch and colleagues.[1 4] It appeared that many studies fail to establish HRQoL and only present mortality rates and crude measures of childhood morbidity. With few exceptions,[5 6] follow up data in adults are lacking. Furthermore, most studies appeared to stress physical rather than social or psychological functioning. To estimate the significance to an individual of an impairment or a functional limitation, generic HRQoL measures—containing physical, mental, and social domains—should be used. These allow for comparisons across patients suffering from various conditions, bringing out the relative severity of diseases.

To fill the knowledge gap, we analysed long term HRQoL effects for congenital anorectal malformations (ARM) and congenital diaphragmatic hernia (CDH). Studies in ARM patients, which predominantly focused on impairments and limitations in specific domains of functioning, suggested a relatively poor long term HRQoL, even after successful surgical reconstruction.[7–9] CDH patients, though often faced with life threatening morbidity during the neonatal period and a variety of symptoms in their first years of life,[10–12] generally seem to lead healthy lives eventually. This study attempts to clarify the HRQoL of survivors of ARM and CDH in a lifetime setting and looks to the question of how it compares with that of the general population.

## METHODS

### Patients

The patient population was comprised of patients treated for ARM or CDH after the year 1969 in the Sophia Children's Hospital, a level III children's hospital serving a referral area of 3.5 million inhabitants. Patients born after the year 1996 were excluded to ensure a minimal period of one year between the patient's birth and the date of investigation. Data were collected from 1997 to 1999. Study questionnaires were distributed by post. If patients did not respond, they were reminded once with a telephone call.

### Outcome measures

The patients or their parents were asked to complete both a symptom checklist created by ourselves and an existing generic HRQoL questionnaire. The former served to measure symptoms. A symptom is defined as a patient's perception of an abnormal physical, emotional, or cognitive state.[13] Patients

**Abbreviations:** ARM, congenital anorectal malformations; CDH, congenital diaphragmatic hernia; ECMO, extracorporeal membrane oxygenation; HRQoL, health related quality of life

aged from 12 years filled in the symptom checklist themselves. Parents filled in proxy versions of the questionnaires on behalf of younger patients.

Because of the rapid developmental change in children, different HRQoL questionnaires had to be used for three age groups. These were the TNO-AZL Infant Quality of Life (TAIQOL) questionnaire (ages 1–4), the TNO-AZL Children's Quality of Life (TACQOL) questionnaire (ages 5–15), and the MOS Short-Form 36 (SF-36) questionnaire (ages 16+). The HRQoL questionnaires were self administered from 16 years on.

### Symptom checklists

The symptom checklist for ARM patients was comprised of seven items, among which were lack of urge sensation, inability to hold faeces, the use of aids for defecation and diapers, and urinary incontinence (see table A, available on the *ADC* website). The total symptom score ranges from 0 to 12, representing maximum symptomatology. The CDH symptom checklist, covering 14 items, dealt with several respiratory difficulties and stomach problems (see table B, available on the *ADC* website). Again, higher scores correspond to more severe symptomatology, with a maximum score of 45.

Clearly, the symptoms studied may also be prevalent in the general population. For example, lack of continence occurs in children born with ARM as well as in healthy children up to a certain age. Therefore, we set out to determine reference scores using a randomly selected sample of children who visited the day-care department of our hospital in 1998 for minor day-case surgery. These controls, who had no record of stool difficulties or respiratory problems, were administered both the ARM checklist and the CDH checklist.

### TAIQOL questionnaire

The TAIQOL questionnaire was developed to be a reliable and valid instrument for measuring HRQoL in children between the ages of 1 and 4.[14] It includes 13 domains: lungs, stomach, skin, sleeping, appetite, eating problems, aggressive behaviour, positive emotions, emotions of anxiety, vitality, social behaviour, motor problems, and communication. The last three domains are applicable only to children aged 18 months and older. The number of items per domain varies from three to seven. Regarding eight of the domains, the TAIQOL investigates HRQoL by assessing functional problems weighted by the degree to which a child shows negative emotions in response to such problems. An example of such a TAIQOL item pair is presented in fig A (see the *ADC* website). In the other five domains, the TAIQOL only measures the frequency of a specific limitation (see fig B on the *ADC* website for an example). Crude domain scores were linearly transformed to a 0–100 scale, with higher scores indicating better HRQoL.

### TACQOL questionnaire

The developers of the TAIQOL also created an instrument for use with children between the ages of 5 and 15 years, named the TACQOL questionnaire.[15] Its seven domains are: pain and symptoms, basic motor functioning, autonomy, cognitive functioning, social functioning, global positive emotional functioning, and global negative emotional functioning. Each domain consists of eight item pairs.[16] The first part of each item assesses the presence of health status problems. The second part assesses the emotional response to such problems. As with the TAIQOL questionnaire, each item pair is encoded into one single score, ranging from 0 to 4. However, no emotional responses are asked regarding ''global positive emotional functioning'' and ''global negative emotional functioning'', since this would lead to nonsensical results. In these two domains, the item scores range from 0 to 2. Consequently, the domain scores range from 0 to 32 for all domains, except those concerning emotional functioning, which range from 0 to 16. Higher scores correspond to better HRQoL. Both the TAIQOL and the TACQOL have been validated in several patient groups since they came into use, with promising results.[16 17]

### SF-36 questionnaire

The SF-36 questionnaire consists of 36 items organised into eight domains: physical functioning, role–physical, bodily pain, general health, vitality, social functioning, role–emotional, and mental health. The SF-36 domain scores range from 0 to 100, with higher scores indicating better HRQoL. Results can be aggregated into a physical and a mental health summary measure. These summary measures were linearly transformed to a mean of 50 and standard deviations of 10 in the general US population. Evidence on the psychometric performance of the widely used SF-36 is mounting.[18 19]

### Statistics

Statistical analyses were carried out using the *t* test for two independent sample means and the *t* test for the significance of a correlation (two-tail probabilities). Results were considered statistically significant if they were at the $p < 0.05$ level.

## RESULTS

### Respondents

Of all 526 ARM patients and 285 CDH patients who underwent treatment for their conditions in the study period, we excluded patients who were deceased (ARM, n = 54; CDH, n = 86), were severely cognitively disabled (ARM, n = 8; CDH, n = 7), or could not be traced (ARM, n = 53; CDH, n = 24). The remaining 411 ARM patients and 168 CDH patients were sent questionnaires. The response rate amounted to 70% (n = 286) in ARM patients and 66% (n = 111) in CDH patients. Because extracorporeal membrane oxygenation (ECMO) was not applied in the hospital until January 1992, only four (all in the 1–4 years age group) of all 111 CDH patients participating in this study were treated with ECMO.

The respondents did not differ from the non-respondents in terms of sex or age. Males comprised a larger proportion of the respondents (59% in ARM patients versus 53% in CDH patients). ARM patients were between 1 and 51 years of age (mean 15.1, SD 9.4). Age ranged from 1 to 42 years in CDH patients (mean 14.3, SD 8.6). There were no differences between ARM and CDH patients and the general population with respect to educational level, involvement in a relationship, or having children.

Symptom checklists were sent to (the caregivers of) 72 children in the control group and returned by 53 (74%).

### Disease specific symptoms

Across all age groups, ARM patients had higher symptom scores than the control group (table 1). Over the years, the symptom score decreased in ARM patients, but remained statistically significantly higher than that of the reference group. To mention a few examples: 68% of all ARM patients sometimes or regularly soiled themselves versus 32% in the control group. Compared to the reference group, a relatively small part of the ARM patients was able to hold faeces for at least one minute (67% *v* 92%) or was able to recognise the type of faeces (64% *v* 85%).

The results presented in table 1 provide fairly strong evidence that, overall, the symptom score in CDH patients was higher than that of the control population. In the control

**Table 1** Symptom scores of ARM patients and CDH patients compared with reference scores

| | ARM patients | | | CDH patients | | | Reference group | | Symptom score difference with ref. group (95% CI†) | |
|---|---|---|---|---|---|---|---|---|---|---|
| Age class | n | Mean age | Mean score (SD) | n | Mean age | Mean score (SD) | n | Mean age | ARM | CDH |
| 1–4 y | 40 | 2.7 | 5.9 (3.3) | 20 | 2.9 | 7.3 (6.1) | 18 | 3.0 | 3.5 (1.7 to 5.3)*** | 1.6 (−2.6 to 5.8) |
| 5–10 y | 67 | 7.2 | 4.2 (3.1) | 23 | 8.0 | 7.5 (7.4) | 22 | 7.3 | 2.9 (1.5 to 4.3)*** | 2.4 (−1.4 to 6.2) |
| 11–15 y | 52 | 12.9 | 3.8 (2.9) | 16 | 13.5 | 5.9 (5.1) | 11 | 12.9 | 3.2 (1.4 to 5.0)*** | 2.1 (−1.4 to 5.6) |
| ⩾16 y | 127 | 23.8 | 2.7 (2.5) | 52 | 21.8 | 7.1 (5.4) | 2 | 17.0 | 2.7‡ | 4.3‡ |
| Total | 286 | 15.0 | 3.7 (3.0) | 111 | 14.3 | 7.0 (5.9) | 53 | 7.4 | 2.2 (1.4 to 3.1)*** | 2.1 (0.2 to 3.9)* |

A missing value analysis (using an expectation-maximisation algorithm) was performed for 79 (16%) of all 503 calculated symptom scores.
†95% confidence interval for the difference between the group means.
‡Difference not tested (because of small sample size).
*Significant at $p < 0.05$.
***Significant at $p < 0.001$.

group, the symptom score showed a clear decrease over the years, which does not match the pattern found in the CDH patients. Many respiratory difficulties and stomach problems were not unique to CDH patients, but almost all symptoms were reported more frequently in CDH patients than in the control group. For example, 76% of all CDH patients never coughed up sputum when waking up, in comparison with 90% in the control group. Of the CDH patients, 77% never experienced constraints as a result of respiratory difficulties, compared to 91% in the control group.

## Health related quality of life

In the youngest age group, differences in HRQoL between patients and the general population were found (table 2). In as many as eight of the 13 TAIQOL domains, the difference between ARM patients and the general population is considered statistically significant. Of note is the low score of the ARM patients on the stomach domain, which consists of the items stomach ache or abdominal pain, colic, and nausea. In most domains, the scores of the CDH patients exceeded those found in the ARM patients. Nevertheless, statistically significant differences between the CDH sample and the reference group were found in five domains. By conventional statistical criteria, we found no clear evidence of a difference in the domain ''lungs'', which is composed of the items bronchitis, difficulties with breathing or lung problems, and shortness of breath.

Tables 3 and 4 present the results of the TACQOL and the SF-36. In ARM patients, four TACQOL domains appeared to discriminate between patients and the general population. The outcomes indicate that the CDH patients scored relatively low in the domains ''basic motor functioning'' and ''cognitive functioning''. Neither in ARM patients nor in CDH patients did the eight domains of the SF-36 or the physical summary measure discriminate between patients and the general population (table 4). The mental summary measure suggests that ARM and CDH patients are even healthier than the general population. In summary, the long term outlook of survivors of these congenital anomalies is favourable. However, there appeared to be some variability in the data, implying that small subgroups of the patients scored relatively low on the HRQoL questionnaires. The poor HRQoL outcomes in these subgroups were associated with the presence of disease specific symptoms. This is illustrated by the negative relation between the SF-36 physical summary score and the symptom score (Pearson's correlation −0.41 in ARM patients ($n = 124$; $p < 0.001$) *v* −0.50 in CDH patients ($n = 50$; $p < 0.001$)). The correlation between the SF-36

**Table 2** TAIQOL scores of ARM patients and CDH patients aged 1–4 years compared with reference scores

| | Mean scores (SD) | | Difference with ref. group† (95% CI‡) | |
|---|---|---|---|---|
| Domain | ARM (n = 41§) | CDH (n = 20) | ARM | CDH |
| Lungs | 91.3 (21.0) | 86.8 (18.7) | −2.2 (−7.7 to 3.4) | −6.6 (−14.1 to 0.9) |
| Stomach | 65.4 (23.8) | 84.2 (22.6) | −26.5 (−31.6 to −21.5)*** | −7.8 (−14.4 to −1.1)* |
| Skin | 91.1 (11.2) | 94.2 (9.8) | −0.9 (−4.4 to 2.6) | 2.2 (−2.5 to 7.0) |
| Sleeping | 73.0 (22.5) | 81.6 (24.6) | −9.0 (−14.9 to −3.1)** | −0.5 (−8.6 to 7.6) |
| Appetite | 76.9 (16.7) | 87.7 (10.9) | −7.5 (−12.0 to −3.1)** | 3.3 (−2.7 to 9.3) |
| Eating problems | 83.1 (15.8) | 91.7 (11.8) | −13.0 (−16.0 to −10.0)*** | −4.5 (−8.2 to −0.7)* |
| Aggressive behaviour | 64.1 (18.2) | 66.8 (19.1) | −3.6 (−8.7 to 1.6) | −0.9 (−8.0 to 6.2) |
| Positive emotions | 93.5 (17.0) | 94.7 (13.7) | −5.2 (−7.9 to −2.4)*** | −3.9 (−7.2 to −0.6)* |
| Emotions of anxiety | 66.7 (21.1) | 76.7 (17.4) | −11.4 (−17.4 to −5.4)*** | −1.4 (−9.5 to 6.8) |
| Vitality | 88.2 (23.0) | 92.5 (19.1) | −9.8 (−13.3 to −6.2)*** | −5.5 (−9.6 to −1.3)** |
| **For children from 18 months of age** | | | | |
| Social behaviour | 88.0 (19.8) | 87.5 (19.4) | −3.0 (−9.5 to 3.5) | −3.5 (−12.3 to 5.3) |
| Motor problems | 92.9 (12.5) | 88.4 (10.4) | −5.8 (−7.6 to −3.9)*** | −10.3 (−12.3 to −8.2)*** |
| Communication | 88.6 (14.5) | 89.8 (13.0) | −3.0 (−6.5 to 0.5) | −1.8 (−6.4 to 2.8) |

†Reference scores (n = 323) were obtained from the creators of the instrument at TNO Prevention and Health, Netherlands (unpublished data). After our investigation, the TAIQOL was replaced by the almost identical TNO-AZL Preschool Children Quality of Life (TAPQOL) questionnaire. Normative data of the TAPQOL, of which an English version is available, have been published.[17]
‡95% confidence interval for the difference between the group means.
§n = 39 for the domains only applicable to children from 18 months of age.
*Significant at $p < 0.05$.
**Significant at $p < 0.01$.
***Significant at $p < 0.001$.

**Table 3** TACQOL scores of ARM patients and CDH patients aged 5–15 years compared with reference scores

| Domain | Mean scores (SD) | | Difference with ref. group† (95% CI‡) | |
|---|---|---|---|---|
| | ARM (n = 118) | CDH (n = 39) | ARM | CDH |
| Pain and symptoms | 27.5 (3.5) | 28.1 (2.9) | −0.1 (−0.8 to 0.6) | 0.5 (−0.6 to 1.7) |
| Basic motor functioning | 30.0 (4.0) | 30.3 (3.0) | −1.0 (−1.5 to −0.6)*** | −0.7 (−1.5 to 0.0)* |
| Autonomy | 30.6 (3.2) | 30.9 (2.5) | −0.8 (−1.1 to −0.4)*** | −0.4 (−1.0 to 0.2) |
| Cognitive functioning | 29.0 (4.8) | 28.3 (4.2) | −0.5 (−1.2 to 0.2) | −1.2 (−2.4 to −0.1)* |
| Social functioning | 29.3 (3.7) | 29.9 (2.3) | −0.7 (−1.2 to −0.3)** | −0.1 (−0.8 to 0.6) |
| Global positive emotional functioning | 14.6 (2.3) | 14.8 (1.8) | −0.4 (−0.7 to 0.0)* | −0.2 (−0.8 to 0.4) |
| Global negative emotional functioning | 11.9 (2.5) | 12.4 (2.3) | 0.2 (−0.3 to 0.6) | 0.7 (0.0 to 1.5) |

The English version of the TACQOL is available on request.
†Reference scores (n = 1311) were derived from a random sample of Dutch children (aged 6–11) in the general population after exclusion of all children with any (parent reported) chronic condition.[16] The means and standard deviations of six age and sex matched groups were combined.
‡95% confidence interval for the difference between the group means.
*Significant at $p < 0.05$.
**Significant at $p < 0.01$.
***Significant at $p < 0.001$.

mental summary score and the symptom score amounted to −0.20 in ARM patients (n = 124; p = 0.02) and −0.47 in CDH patients (n = 50; p = 0.001).

## DISCUSSION

In this paper, we have been concerned with the HRQoL of survivors of ARM and CDH in a long term setting. Many of them appeared to retain substantial residual symptomatology. Compared with reference data, the HRQoL of the patients (especially those with ARM) aged 1–4 was poor. In the patients aged 16 years and over, hardly any differences with the general population were found. It must be stressed that the relatively poor outcomes in the youngest patients are not offset by the encouraging outcomes in adult life. This stage of life has its own inherent importance for the developing individual. Therefore, future research should especially be targeted at finding ways to improve the HRQoL of the youngest children. It is clear from the current study that the relations between symptoms, impairments, limitations in functioning, and HRQoL should be interpreted with caution. An important lesson to be learned is that impairments are imperfect predictors of HRQoL. Not every impairment automatically triggers a decrease of the HRQoL.

The HRQoL of the ARM patients aged between 1 and 4 years was statistically significantly lower than population standards predicted, even though complaints of incontinence are not prominent until patients grow older. Their HRQoL improved considerably with growing age, notwithstanding the fact that stool difficulties are widely believed to disrupt HRQoL due to pain, feelings of shame, or inability to take part in social activities such as sports. A possible explanation is that diapers, therapeutic aids such as enemas, and dietary manipulations are effective tools in restoring satisfying functioning. Seemingly, the patients are able to cope successfully with their handicaps in one way or another.

A HRQoL questionnaire did not reveal a pronounced difference in the lung domain between CDH patients aged 1–4 years and a reference group. In a sense, these results are consistent with earlier research we performed in this specific patient group (median age 11.7 years) using lung function tests.[12] This study showed that in the long term no severe pulmonary impairment resulted from CDH. The reduced level of cognitive functioning in the CDH patients aged 5–15 years (none treated with ECMO) corroborates a small scale study previously carried out in our hospital.[20] In the current study, from a certain age the HRQoL of the CDH patients could hardly be distinguished from that of the general population. One explanation is that, particularly in the past, a major ''selection'' took place in the first week of life due to the high mortality in patients with severe pulmonary hypoplasia and therapy resistant pulmonary hypertension. In other words, those patients in the worst shape are likely not to have survived. Finally, it should be noted that, while ECMO improves survival in selected, critically ill infants with CDH,[21] it is also true that the ECMO technique enables us to keep some patients alive with a relatively poor

**Table 4** SF-36 scores of ARM patients and CDH patients aged 16 years and older compared with reference scores

| Domain | Mean scores (SD) | | Difference with ref. group† (95% CI‡) | |
|---|---|---|---|---|
| | ARM (n = 127) | CDH (n = 52) | ARM | CDH |
| Physical functioning | 92.8 (14.3) | 93.7 (14.0) | −0.3 (−2.7 to 2.1) | 0.6 (−2.8 to 4.0) |
| Role–physical | 87.6 (28.5) | 90.2 (28.8) | 1.2 (−4.2 to 6.6) | 3.8 (−4.1 to 11.7) |
| Bodily pain | 78.2 (17.0) | 83.5 (11.4) | −2.7 (−6.3 to 1.0) | 2.6 (−2.8 to 8.0) |
| General health | 75.3 (22.1) | 78.5 (17.7) | −2.9 (−6.4 to 0.7) | 0.3 (−4.7 to 5.2) |
| Vitality | 68.0 (17.2) | 68.1 (21.7) | −2.7 (−5.9 to 0.5) | −2.6 (−7.4 to 2.2) |
| Social functioning | 89.9 (15.6) | 91.1 (20.3) | 2.1 (−1.5 to 5.6) | 3.3 (−2.2 to 8.8) |
| Role–emotional | 90.1 (26.8) | 90.9 (28.3) | 4.7 (−1.0 to 10.4) | 5.4 (−3.1 to 14.0) |
| Mental health | 78.3 (15.1) | 77.4 (19.4) | −0.4 (−3.4 to 2.5) | −1.3 (−5.8 to 3.1) |
| Physical component summary | 52.2 (7.7) | 53.7 (5.2) | −0.7 (−2.1 to 0.7) | 0.9 (−1.2 to 3.0) |
| Mental component summary | 52.5 (8.0) | 51.9 (10.9) | 3.6 (1.8 to 5.3)*** | 3.0 (0.3 to 5.7)* |

Details of the SF-36 are available at http://www.sf-36.org/demos/SF-36.html.
†Reference scores for the eight ''individual'' domains were derived from a Dutch general population sample, aged 16–40. ''n'' was estimated at 551 on the basis of the fact that the total sample, aged 16–94, contained 1742 people.[34] To obtain reference scores for the summary scales, we had to rely on a US general population sample, aged 18–44 (n = 2765).[35]
‡95% confidence interval for the difference between the group means.
*Significant at $p < 0.05$.
***Significant at $p < 0.001$.

prognosis and that it may lead to iatrogenic morbidity.[22] Thus, close follow up of CDH patients becomes even more essential in the future.

When evaluating the outcome of ARM and CDH, we did not include the patients who had died. Although it is of major importance to critically evaluate the deaths—as was done earlier in our paediatric surgical department[23 24]—this was beyond the scope of the current study. Here, we concentrated on assessing whether improved survival in neonatal surgery comes at the expense of poor HRQoL. Apart from that, 78% of the 54 deceased ARM patients died within their first year of life, with a median life span of 23 days. Of the 86 CDH patients who died, 87% died within their first year of life (median life span 3 days). The vast majority of the deceased were therefore too young to assess the quality of their lives. Similarly, we did not assess patients who were severely cognitively disabled. Cognitive disability, which is attributable to a wide variety of underlying causes ranging from birth asphyxia to severe chromosomal anomalies (including trisomy 21), is not common in these patient groups. Only 1.8% of all eligible patients had to be excluded for this reason (n = 15; mean age 24.6 years). A solid investigation into the HRQoL of this subgroup would have been difficult to accomplish within the current study. HRQoL can be assessed by proxy in these subjects, but one must seriously doubt whether the measures used would be sufficiently valid and reliable in these cases. The severely cognitively disabled patients may differ in their perception of HRQoL, and their ability to express their experiences is often very limited. This may reduce the parents' or caregivers' ability to make accurate judgements about aspects of the patient's HRQoL.

Assessment of HRQoL in children poses special problems. Because of difficulties that small children have with notions of abstract concepts and language, we had to rely on proxies. We are conscious of the variety of factors that can influence a parent's rating of his or her child's HRQoL and the equivocal findings reported in the literature.[25–27] Nevertheless, recent research findings in children[28–30] suggest that a parent is able to report appropriate information regarding his or her child's HRQoL, especially concerning observable behaviours. There is as yet no clear evidence of whether—when the parent and the child disagree—the parents over- or under-estimate HRQoL. A number of studies indicated that parents tend to rate the child as having a poorer HRQoL than the child does him or herself, a tendency which would result in a conservative estimate of the HRQoL.[31–33] These studies, often concentrating on acquired conditions, apply, however to children aged between about 7 and 13, who are able to give self reports, while our study also comprised younger children. Finally, as in most previous studies in this area, we limited ourselves to one source of information and did not obtain additional views of, for example, other relatives or teachers. Yet, the use of proxies other than parents may have value and merits further investigation.

In conclusion, our investigation into the HRQoL of survivors of ARM and CDH provides favourable results. The suggestion that improved survival can only be reached at the price of poor HRQoL is not substantiated. As expected a priori, ARM and CDH can cause considerable symptomatology. Nevertheless, the vast majority of the patients ultimately enjoy healthy lives.

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# ARCHIVIST

## Pathologists' terminology for sudden infant deaths

Whatever terms are used for sudden unexpected deaths in infancy (SUDI) it is important that terminology should be agreed and standardised; everybody should "sing from the same hymn sheet". A survey carried out for the Foundation for the study of Infant Deaths, in collaboration with the Office of National Statistics and the Royal College of Pathologists (S Limerick and C Bacon. *J Clin Pathol* 2004;**57**:309–11) has shown that pathologists vary in their use of terms when reporting on SUDI.

A questionnaire was sent to all 105 pathologists known by the Royal College of Pathologists to perform SUDI autopsies. There were 63 satisfactory replies (five responders said they no longer did SUDI autopsies and there were 37 nonresponders). Of the 63, 29 were forensic pathologists, 24 paediatric pathologists, and 10 general pathologists (six with forensic accreditation). They were asked if they used the term "unascertained" when they found no adequate cause for death. Eleven always used the term, 19 used it frequently, 26 occasionally, and seven never. Of the 56 pathologists who used the term 38 would use it whenever death had occurred while the infant was sharing a bed with an adult and 37 when there were suspicious features in the history or at autopsy. Eight pathologists admitted using the term to obtain an inquest.

Sixty-two respondents answered a question about information available to them at the time of the autopsy and half (32) of these complained that the information was usually inadequate; they wanted more information about the medical and social history and the antecedents and circumstances of death. Fifty-two routinely received a report from the coroner's officer but only ten regularly had a report from a health professional. Thirty-two said they sometimes visited the scene of the death. Thirty-eight pathologists believed that the term SIDS was still useful, 24 did not, and one was undecided. For sudden unexplained death while sharing a bed with an adult 38 would use the term unascertained, 25 would call it SIDS, six SUDI, and one asphyxia (some would use different terms in different circumstances). Eighteen of the 63 pathologists performed fewer than six SUDI autopsies a year.

There is variation in the use of the term "unascertained" but whereas SIDS implies a natural cause, unascertained does not exclude unnatural death. The authors of this paper point out that use of the term "unascertained" could therefore stigmatise the family. They call for all SUDI to be given the same designation initially and for more specific, or potentially accusatory, terms to be used only after full investigation, often including an inquest.