Long-term prognosis for children with breath-holding spells

Anne Lise Olsen¹, Rene Mathiasen², Niels Henrik Rasmussen¹ & Finn Ursin Knudsen²

ABSTRACT
INTRODUCTION: The aim of this study was to investigate the long-term prognosis for children with severe breath-holding spells (BHS).

MATERIAL AND METHODS: The study was a retrospective cohort study. Data from the medical records of all patients with severe BHS admitted during a ten-year period were recorded and a questionnaire was sent to the families. A matched group of adolescents with febrile convulsions served as controls (n = 289).

RESULTS: A total of 85 out of 115 families (73.9%) responded. The mean age of the included patients was 20.5 years. Among first-degree relatives 21.5% had BHS and 14.6% had epilepsy. The peak age for severe BHS was 16 months. The children had a total of 1-25 attacks. All electrocardiogram (ECG) recordings were normal. One child died of asystolia at the age of 20 years, ECG two weeks previously showed a WPW-block. Twenty-six (30.6%, p < 0.001) had fainting spells. Twenty-five (29.4%) had concentration problems.

The grades achieved by BHD children at the final school exam did not differ from the mean values achieved by all children in the area.

CONCLUSIONS: In this study on the long-term prognosis of children with BHS, we found a predisposition to fainting spells as expected. We also found that 29.4% of children with BHS had concentration problems. Further follow-up studies are needed to confirm this trend.

Breath-holding spells (BHS) is a clinical entity that is easy to diagnose in its typical form. There are two clinical subtypes of breath-holding spells, which are based on the child’s colour – the cyanotic (blue) and the pallid type [1, 2]. Severe BHS causes unconsciousness. About 4% of all children under the age of five years are affected [1].

Breath-holding spells usually resolve spontaneously with no long-term sequelae. In the literature it is generally agreed that the prognosis for breath-holding attacks is excellent, long-term prognosis is considered good and it is expected that the children will develop normally, except for an increased risk of developing syncope later in life. No other lasting sequelae of BHS have been described, but only a few long-term studies were published [1, 3, 4].
were compared with the mean values for the population of the area.

Data collection took place between February 1 2003 and July 1 2003. The mean age of the children at the time of data collection was 20.5 years (range = 14.7-27.9). Children from our previous study on febrile convulsions [5] served as controls, as they had a similar age distribution, were recruited from the same area of Copenhagen, and were admitted to the same paediatric departments. It is well documented that children with febrile convulsion have an excellent prognosis and are comparable with children in general [5]. When available, data for the background population in the area were used for further comparison. Data were analysed using Pearson’s χ² test.

The local ethical committee approved the study.

RESULTS

A total of 125 patients were admitted with the diagnosis of BHS during the ten-year period covered by the study. Ten patients were excluded due to emigration (n = 2), unknown home address (n = 2), unconfirmed diagnosis (n = 2), death during follow-up (n = 2, see below), psychomotor retardation (n = 2) (neurofibromatosis I (n = 1), shaken baby syndrome with subdural haematoma and retinal bleedings (n = 1).

Two patients died during follow-up, both had pallid spells, none of them had an ECG performed during the time they were admitted to hospital for BHS. One died in a traffic accident unrelated to BHS, and the other due to ECG-verified asystoly at the age of 20 years, following two months of near-fainting attacks. This patient suffered no verified cardiac problems during childhood or adolescence. Resuscitation attempts in the outpatient clinic were unsuccessful. ECG two weeks previously revealed a Wolf-Parkinson-White (WPW)-block with a short PQ-interval, slightly broadened QRS-complexes and delta waves in the standard recordings.

Among the 115 questionnaires sent, 85 families (73.9%) to 34 girls and 51 boys responded. The mean birth weight was 3,212 g, (range: 1,500-4,400 g), (n = 78), the mean birth length was 50 cm (range: 44-56 cm), (n = 70), mean gestational age was 39 weeks (range: 30-43 weeks), (n = 50). The proportion with perinatal problems was 31.1% versus 25.9% in the controls, and did not differ from the values observed in the normal population. In the BHS group 17 first-degree relatives (21.5%) had BHS and 12 (14.6%) had epilepsy, i.e. much higher incidences than in the background population (Table 1).

A total of 38 (44.7%) had blue BHS only, 13 (15.3%) had pallid BHS only, nine (10.6%) had the mixed form and 12 (14.6%) had epilepsy, i.e. much higher incidences than in the background population (Table 1).

Epilepsy in first degree relatives 12/82 (14.6) 15/269 (5.2) 0.023

Severely abnormal 3/6 (48) 7/262 (2.7) NS

Slightly/moderately abnormal 7/6 (11.2) 26/262 (9.9) NS

Abnormal electroencephalography 10/62 (11.2) 33/262 (12.5) < 0.001

Delayed psychomotor development 1/72 (1.4) 11/274 (4.0) NS

Perinatal problems 23/73 (31.5) 73/287 (25.4) NS

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Epilepsy in first degree relatives 12/82 (14.6) 15/269 (5.2) 0.023

Febrile convulsion in first degree relatives 2/85 (2.4) –

TABLE 1

Epidemiological data for children with breath-holding spells and a control group (comprising children with febrile convulsions).

<table>
<thead>
<tr>
<th>BHS</th>
<th>Controls</th>
</tr>
</thead>
<tbody>
<tr>
<td>n/N (%)</td>
<td>n/N (%)</td>
</tr>
<tr>
<td>Perinatal problems</td>
<td>23/73 (31.5)</td>
</tr>
<tr>
<td>Delayed psychomotor</td>
<td>1/72 (1.4)</td>
</tr>
<tr>
<td>Abnormal electroence</td>
<td>10/6 (16.1)</td>
</tr>
<tr>
<td>Slightly/moderately</td>
<td>7/6 (11.2)</td>
</tr>
<tr>
<td>Severe abnormal</td>
<td>3/6 (4.8)</td>
</tr>
<tr>
<td>BHS in first degree</td>
<td>17/79 (21.5)</td>
</tr>
<tr>
<td>Epilepsy in first</td>
<td>12/82 (14.6)</td>
</tr>
<tr>
<td>degree relatives</td>
<td></td>
</tr>
<tr>
<td>Febrile convulsion</td>
<td>2/85 (2.4)</td>
</tr>
</tbody>
</table>

BHS = breath-holding spells; NS = not significant

(a) Number of patients with outcome variable/total number of patients.

TABLE 2

Long-term outcome parameters for children with breath-holding spells and a control group (comprising children with febrile convulsions).

<table>
<thead>
<tr>
<th>BHS</th>
<th>Controls</th>
</tr>
</thead>
<tbody>
<tr>
<td>n/N (%)</td>
<td>n/N (%)</td>
</tr>
<tr>
<td>Febrile convulsions</td>
<td>6/85 (7.1)</td>
</tr>
<tr>
<td>Epilepsy</td>
<td>0/84 (0.0)</td>
</tr>
<tr>
<td>Fainting spells</td>
<td>26/85 (30.0)</td>
</tr>
<tr>
<td>Migraine</td>
<td>9/84 (10.7)</td>
</tr>
<tr>
<td>Tension type headache</td>
<td>25/85 (29.4)</td>
</tr>
<tr>
<td>Minor motor problems</td>
<td>10/85 (11.8)</td>
</tr>
<tr>
<td>Specialized education</td>
<td>26/85 (31.7)</td>
</tr>
<tr>
<td>Concentration problems</td>
<td>25/85 (29.4)</td>
</tr>
</tbody>
</table>

BHS = breath-holding spells; NS = not significant

(a) Number of patients with outcome variable/total number of patients; (b) Incidence in the Danish childhood population [6].
In the BHS group, the mean grade achieved at the official final exam after attending school for nine years (using a scale ranging from 0 to 13) was 8.11 (range: 6.70-10.90) (n = 44), this did not differ significantly from the mean value for the normal population 8.18, range not available, (n = 14,713).

After ten years of schooling, the official final exam grades of the children with BHS (mean 8.05, range 6.50-10.40) were not different from the mean values observed in the whole area (mean 7.99, range not available, n = 2,461).

In nine (10.6%), academic performance was reduced and 26 (31.7%) needed to attend specialized education classes (Table 2).

The parents of 25 children (29.4%), 13 girls (38% of all girls) and 12 boys (23% of all boys), confirmed that their child had experienced concentration problems.

The distribution of BHS types did not differ in this group compared to the group without concentration problems. A total of 48% had blue BHS only versus 43% in the group without concentration problems; 16% versus 15% had pallid BHS only; 16% versus 5% had the mixed form; and 20% versus 33% could not be classified.

Comparing the number of attacks in the two groups, we found that 28% of the children in the group with reported concentration problems had one attack versus 30% in the group without concentration problems; 12% versus 10% had two attacks; eight versus seven percent had three attacks; 28% versus 43% had 4-7 attacks and 24% versus 10% had more than eight attacks.

**DISCUSSION**

We found a similar distribution between pallid, blue, mixed and not classified spells to that reported in previous studies [1, 3, 4]. There were no significant differences in perinatal data when comparing the BHS group to a normal birth-cohort or to the control group. No significant perinatal problems were reported.

It has been suggested, that iron deficiency may play a role in the pathogenesis of BHS [7, 8] but in this study 25 children were examined, 23 of whom had a normal haemoglobin level. The patient who died aged 20 from undiagnosed WPW, draws attention to the importance of performing an ECG, at least in all children with pallid BSH, in order to identify prolonged QT-syndrome or other cardiac arrhythmia [2].

Our results confirm the previously established familial tendency to BHS [1, 4]. We also found a familial tendency to epilepsy (Table 2), which has not been described in previous follow-up studies.

The long-term outcome in this study confirms the predisposition to fainting spells [1, 3, 4]; there were 26 children (30.6% in the BHS group versus 6.3% of the controls, p < 0.001) who later developed fainting spells.

In a prospective study performed in 2001, DiMario found that four of 95 children with BHS were in special education classes [4]. Haverkamp & Noeker evaluated cognitive performance in 28 patients, and found no significant difference in cognitive performance between patients and siblings [9].

32% of the children in our study needed specialized education. Nevertheless, they achieved a normal exam result, which may be the result of the special education received, or a result of cohort selection. Although in our experience most of the children with severe BHS are admitted to hospital. We have not been able to compare the need for specialized education in our population, with the need in an unselected population, but a contemporary Danish study has shown similar frequencies among children in the same area who were referred to a school-psychologist; 29.7% of these children received specialized education during nine years of school attendance [10].

Lombroso & Lerman and Laxdal et al stated that “breath-holders” are frequently stubborn, disobedient, aggressive, hyperactive and irritable [1, 3].

We found that 29.4% of the children had concentration problems. We did not reveal considerable differences when comparing the types of BHS found in this group to those found in the group without concentration problems. The proportion of children who had more than eight attacks in the group with concentration prob-
lems was slightly higher than in the other group, but when we compared children who had more than four attacks, proportions were similar 52% versus 53%.

We did not ask specifically about attention deficit hyperactive disorder, and we did not use a specific rating-scale, and this may be considered limitations of this study.

CONCLUSION
In this study on the long-term prognosis of children with BHS we found a predisposition to fainting spells as expected. We also found a familial tendency to epilepsy, which was not described in previous follow-up studies.

Furthermore, almost a third (29.4%) of the children with BHS reported that they had concentration problems. We were unable to find any differences between the types and frequencies of attacks in the children with concentration problems compared with the children who did not report concentration problems. Further follow-up studies are needed to evaluate our findings.

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CONFLICTS OF INTEREST: None

LITERATURE