CONGENITAL BRACHIAL PALSY: INCIDENCE, CAUSES, AND OUTCOME IN THE UNITED KINGDOM AND REPUBLIC OF IRELAND

G Evans-Jones, S P J Kay, A M Weindling, G Cranney, A Ward, A Bradshaw, C Herron

Objectives: To determine the incidence and study the causes and outcome of congenital brachial palsy (CBP).

Design: Active surveillance of newborn infants using the British Paediatric Surveillance Unit notification system and follow up study of outcome at 6 months of age.

Setting: The United Kingdom and Republic of Ireland.

Participants: Newborn infants presenting with a flaccid paresis of the arm (usually one, rarely both) born between April 1998 and March 1999.

Main outcome measures: Extent of the lesion at birth and degree of recovery at 6 months of age.

Findings: There were 323 confirmed cases giving an incidence of 0.42 per 1000 live births (1 in 2300). Significant associated risk factors in comparison with the normal population were shoulder dystocia (60% v 0.3%), high birth weight with 53% infants weighing more than the 90th centile, and assisted delivery (relative risk (RR) 3.4, 95% confidence interval (CI) 2.9 to 3.9, p = 0.0001). There was a considerably lower risk of CBP in infants delivered by caesarean section (RR 7, 95% CI 2 to 56, p = 0.002). At about 6 months of age, about half of the infants had recovered fully, but the remainder showed incomplete recovery including 2% with no recovery. The relative risk of partial or no recovery in infants with extensive lesions soon after birth compared with those with less extensive lesions was 11.28 (95% CI 2.38 to 63.66, p = 0.000005).

Conclusions: The incidence of CBP in the United Kingdom and Republic of Ireland is strikingly similar to that previously reported nearly 40 years ago. Most cases are due to trauma at delivery, which is not necessarily excessive or inappropriate. Given the uncertainty about the appropriate management of these infants, serious consideration should be given to a formal clinical trial of microsurgical nerve repair.

RESULTS

A total of 22,155 cards were circulated in the 12 month study period; 20,663 (93%) cards were returned. These alerted us to 430 cases of suspected CBP. Questionnaires were sent to all respondents, and 391 (91%) of the first questionnaires were returned. There were 68 ineligible cases (40 because the infants were born outside the study period, 24 duplications, and four cases of revised diagnosis) leaving 323 confirmed cases (table 1). No cases were reported to ONS and ICS that correspond to total "flail arm", the two differing only in the presence of Horner’s syndrome in Group IV.

Ethical approval was obtained from the South Cheshire local research and ethical committee. Statistical analysis was by the χ² test. The study period was April 1998 to March 1999.

METHODS

Active surveillance for CBP was undertaken by the monthly reporting card system of the British Paediatric Surveillance Unit, which invites consultant paediatricians in the United Kingdom and Republic of Ireland who are members of the Royal College of Paediatrics and Child Health to report cases in up to 12 diagnostic groups. We were informed of each case notified, and sent two questionnaires to each respondent requesting information. In the first, we requested information about the obstetric history and clinical findings in the baby, and in the second, information on muscle function in the affected arm at 6 months of age. Complementary sources comprised reports from plastic and orthopaedic surgeons. Results were validated by conducting a search for cases notified to the Office of National Statistics (ONS) and the Irish Census Office (ICS). Each case was classified according to Narakas’s classification, which reflects the clinical findings in the first weeks of life. Narakas’s Group I (C5–C6 lesion) and Group II cases (C5–C7) correspond to Erb’s palsy, with the latter adopting the classical “waiter’s tip” posture because of loss of wrist extension. Narakas’s Groups III and IV (C5–C6 lesion) and Group II cases (C5–C7) correspond to total “flail arm”, the two differing only in the presence of Horner’s syndrome in Group IV.

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but the Irish figure (53 524) is for the exact study period obtained from the ICS.

Male sex was slightly more common (53%) mirroring the slight male preponderance in the normal population.\(^5\)

The right arm was more commonly affected than the left (50% v 43%, with 4.5% not specified), as has been reported previously.\(^6\)

There were five bilateral cases; the right arm was more severely affected than the left in three cases and the reverse in the remaining two. In three of these five cases, the infant was “large for dates” and recognised to have shoulder dystocia. Of the remaining two infants of normal birth weight, one was delivered by ventouse and the other by breech extraction.

Table 2 shows the distribution of mode of delivery in the study population compared with the normal population in England in 1994–1995.\(^5\) The chance of infants who developed CBP being delivered by assisted delivery (ventouse or forceps) (36%) is significantly increased (relative risk (RR) 3.4, 95% confidence interval (CI) 2.9 to 3.9, p = 0.0001) over that in the normal population (10.6%). Infants with CBP also had an increased chance of being delivered by ventouse alone (RR 4.4, 95% CI 1.8 to 10.9, p = 0.001). The chance of being delivered by forceps alone was not significantly increased (RR 2.1, 95% CI 0.9 to 4.8, p = 0.17). The incidence of breech delivery (3%) was not significantly higher than for the general population (1%). Of the five caesarean section deliveries in the study group, one was elective, the remainder emergencies. Compared with the general population\(^5\) (15.5%), considerably fewer infants who developed CBP were delivered by caesarean section (1.5%) (RR 7, 95% CI 2 to 56, p = 0.002).

Shoulder dystocia, defined as difficulty in delivering the shoulders after the head has delivered, occurred in 206 cases (64%) as opposed to an “accepted conservative figure” of 0.3% in the normal population.\(^7\)

![Figure 1](http://fn.bmj.com/)

**Figure 1** Birth weight distribution.
Table 5 Other probable traumatic cases

<table>
<thead>
<tr>
<th>Birth weight (g)</th>
<th>Maternal history</th>
<th>Delivery</th>
</tr>
</thead>
<tbody>
<tr>
<td>3430</td>
<td>Primigravida. Concealed pregnancy</td>
<td>Born at home</td>
</tr>
<tr>
<td>3946</td>
<td>Primigravida. Concealed pregnancy</td>
<td>Born at home in toilet</td>
</tr>
<tr>
<td>3780</td>
<td>Primigravida</td>
<td>Prolonged labour with fetal distress</td>
</tr>
<tr>
<td>3730</td>
<td>Previous delivery; shoulder dystocia. Known cephalo-pelvic disproportion</td>
<td>Prolonged labour</td>
</tr>
<tr>
<td>3880</td>
<td>Previous delivery; shoulder dystocia</td>
<td>Normal vertex delivery</td>
</tr>
<tr>
<td>3540</td>
<td>Primigravida</td>
<td>Prolonged labour</td>
</tr>
<tr>
<td>3084</td>
<td>Primigravida. Failure to progress</td>
<td>Emergency caesarean section</td>
</tr>
<tr>
<td>3500</td>
<td>Failure to progress</td>
<td>Emergency caesarean section (infant had torticolis)</td>
</tr>
<tr>
<td>3560</td>
<td>Hand presentation</td>
<td>Emergency caesarean section with extended incision and cephalic version</td>
</tr>
<tr>
<td>3500</td>
<td>Breech presentation; external cephalic version</td>
<td>Normal vertex delivery at term</td>
</tr>
</tbody>
</table>

Table 6 Details of outcome

<table>
<thead>
<tr>
<th>Number</th>
<th>Full recovery</th>
<th>Partial recovery</th>
<th>No recovery</th>
</tr>
</thead>
<tbody>
<tr>
<td>All infants</td>
<td>276</td>
<td>143</td>
<td>127</td>
</tr>
<tr>
<td>Macrosomic</td>
<td>146</td>
<td>65</td>
<td>78</td>
</tr>
<tr>
<td>Shoulder dystocia</td>
<td>179</td>
<td>64</td>
<td>90</td>
</tr>
<tr>
<td>Spontaneous vertex delivery</td>
<td>156</td>
<td>80</td>
<td>74</td>
</tr>
<tr>
<td>Breech</td>
<td>19</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>Assisted delivery</td>
<td>101</td>
<td>32</td>
<td>44</td>
</tr>
<tr>
<td>Ventrine</td>
<td>80</td>
<td>40</td>
<td>39</td>
</tr>
<tr>
<td>Forceps</td>
<td>37</td>
<td>24</td>
<td>12</td>
</tr>
<tr>
<td>Caesarean section</td>
<td>3</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Groups I and II</td>
<td>255</td>
<td>143</td>
<td>112</td>
</tr>
<tr>
<td>Groups III and IV</td>
<td>19*</td>
<td>0</td>
<td>13</td>
</tr>
<tr>
<td>Associated injuries</td>
<td>42</td>
<td>20</td>
<td>17</td>
</tr>
<tr>
<td>Age at assessment (weeks)</td>
<td>Range: 1–70</td>
<td>1–75</td>
<td>19–33</td>
</tr>
<tr>
<td>Median</td>
<td>18</td>
<td>26</td>
<td>27</td>
</tr>
</tbody>
</table>

Notes: Completed questionnaires = 281. Insufficient information on outcome or ineligible cases = 5. Total number of eligible cases = 276. Information on the mode of delivery was missing in two fully recovered and five partially recovered infants. The sum of ventouse and forceps deliveries exceeds the number of infants delivered by assisted delivery because some proceeded from ventouse to forceps. *Information on the extent of the lesion was missing in two cases.

Table 3 and figure 1 show the distribution of birth weights in the study group as compared with singletons for 1994–1995 in England,1 with a shift to the right. The range was 600–5840 g with a median of 4180 g compared with 3380 g in the normal population.2 A total of 170 infants (53%) weighed more than the 90th centile at term for the normal population (4065 g)—that is, were “large for dates” or “macrosomic”—based on the Gairdner-Pearson growth charts first published in 1971 and revised in 1985 (Castlemead Publications).

Of the 323 cases reported, 293 (91%) were in Narakas’s Groups I and II, 21 (6.5%) in Groups III and IV, with information missing in the remainder. The five bilateral cases were classified according to the most severely affected arm. No cases of “Klumpke’s palsy” affecting C8 and T1 only were reported.

Table 4 summarises the occurrence of associated injuries. There were 26 (8%) bony injuries, the most common being a fractured clavicle (11 cases); five cases of Horner’s syndrome were reported, three facial nerve palsies, but only one phrenic nerve palsy, although information on the latter was unavailable or not known in a third of cases. Of other associated injuries, head and other upper body bruising were the most often reported.

In 29 (9%) infants, there was no obvious explanation and no identifiable indicators of the application of unusual force in the course of the delivery—that is, they were not delivered by assisted or breech delivery, did not have shoulder dystocia or other injuries, and were not macrosomic. However, when further details were obtained about infants in this group, probable factors leading to brachial plexus trauma were identified in 10 cases (table 5), including three infants born by emergency caesarean section. One infant with the breech presenting had external cephalic version at 32 weeks gestation and was delivered normally at term weighing 3500 g. If these cases are regarded as “explained”, then the number for which there is no obvious explanation is reduced to 19 (5.9%).

In one case that fulfilled the case definition, delivery was by elective caesarean section at 32 weeks for growth retardation. Ultrasound scans were performed at 19, 23, and 32 weeks. At 19 and 23 weeks, the left wrist appeared hyperflexed, and at 32 weeks the left arm was also noted to be shorter than the right. At birth, there was a flaccid paresis of the left arm which was generally smaller than the right, with shoulder muscle wasting and a flexion contracture of the left wrist. We are unaware of any reports of a similar case, and the final diagnosis remains unknown.

Of the 323 eligible cases, 281 (87%) second questionnaires were returned; the case described above was excluded and four questionnaires were incomplete, leaving 276 for analysis. The median (range) age of assessment was 23 (18–27) weeks. Figure 2 and table 6 summarise the details of the outcome at follow up. There were 143 (52%) cases with full recovery, 127 (46%) with partial recovery (defined as failure to recover full normal function in any muscle group in the affected arm), and six (2%) with no recovery. Of 146 (53%) macrosomic infants, 65 (45%) made a full recovery, 78 (53%) a partial recovery, and in three (2%) there was no recovery. For the infants who were not macrosomic, the comparative proportions were 60%, 38%, and 2% respectively. For macrosomic infants, the relative risk of no or partial recovery compared with those who were not macrosomic was 1.37 (95% CI 0.91 to 2.04, p = 0.144). A total of 179 (65%) infants had shoulder dystocia. Of these, 84 (47%) showed full recovery, 90 (50%) partial, and five (3%) no recovery compared with 61%, 38%, and 1%, respectively in those without dystocia. The relative risk of no or partial recovery for infants with shoulder dystocia compared with those without was 1.10 (95% CI 0.91 to 1.32, p = 0.336). Of 101 (37%) infants born by assisted delivery, 55 (54%) made a full recovery, 44 (44%) a partial recovery, and two (2%) no recovery compared with 51%, 47%, and 2% respectively in 156 infants born by spontaneous vertex delivery. There was no significantly increased risk for infants delivered by assisted delivery therefore of partial or no recovery compared with infants delivered normally. Of 255 infants with Group I and II lesions, 143 (56%) made a full recovery, 112 (44%) a partial recovery, with none showing no recovery. The comparative figures for 19 Group III and IV cases were 0 (0%), 13 (68%), and six (32%), so that the relative risk for infants with the more extensive lesions of partial or no recovery versus full recovery was 11.28 (95% CI 2.38 to 63.66, p = 0.000005). Table 6 summarises the outcome for infants born by breech extraction and caesarean section and those with associated injuries.
Figures 2 and 3 report the pattern of recovery in infants who showed only partial recovery by 6 months, by consideration of movements solely innervated by roots C5, C6, and C7, namely shoulder abduction, elbow flexion, and elbow extension respectively. This shows the expected centripetal progression of recovery. At 6 months, for example, 68 (54%) infants with incomplete recovery had no or partial recovery of elbow flexion.

DISCUSSION

The current incidence of CBP in the United Kingdom and Republic of Ireland was unknown until this report, possibly the largest ever completed, with 776,618 live births surveyed. The survey was prompted by the development of microsurgical nerve repair procedures for CBP pioneered in adults with traumatic brachial plexus lesions and the need to identify more effectively those infants who may benefit from such intervention. Other factors were an awareness of the impact of CBP on healthcare costs through significant lifelong disability and litigation.

Adler and Patterson in New York reported a reduction in the incidence of CBP from 1.56 per 1000 live births in 1938 to 0.38 per 1000 in 1962, which they ascribed to improved obstetric practice. Subsequent reports have suggested an increase in incidence, possibly because of an increase in birth weight. Bennett and Harrold reported an incidence of 0.61 per 1000 in the United Kingdom in 1976, Greenwald et al reported an incidence of 2.0 per 1000 in the United States in 1984, Sjoberg et al reported an incidence of 1.9 per 1000 in Sweden in 1988, and in 1990 al-Rajeh et al reported an incidence of 1.19 per 1000 in Saudi Arabia.

Our reported incidence of 0.42 per 1000 in the United Kingdom and Republic of Ireland is strikingly similar to that reported in the United States in 1962 in New York. This is in spite of an increase in rates of caesarean section and a fall in the number of vaginal breech deliveries in recent years, a possible reason being the concurrent increase in birth weight. The most important factors associated with the occurrence of CBP, which have been reported previously, were shoulder dystocia, assisted delivery, and high birth weight. The significantly lower incidence than normal of caesarean section in infants with CBP has also been reported previously. The previously reported high incidence of CBP and of extensive lesions in breech delivery was not found in our survey, but only 9 infants were breech deliveries.

The 19 (5.9%) cases in which there was no obvious predisposing factor raises the possibility of non-traumatic causes, although the proportion is much smaller than that reported by Jennet et al, who found an incidence of 56% of unexplained cases in a much smaller survey than this, leading them to propose “intrauterine maladaptation” as the cause. Other factors that could predispose to brachial plexus injury include fetal malposition, cephalopelvic disproportion, dystocia of the posterior shoulder on the sacral promontory, or a combination of these, and have been postulated previously.

CBP should therefore not be regarded as “prima facie” evidence of excessive applied traction and can occur in any part of the head. We are unaware of any previous reports of CBP occurring after external cephalic version.

The most important differential diagnosis in CBP is injury around the shoulder joint causing pseudoaparesis but, in such cases, passive movement is as affected as active movement. Other differential diagnoses include cervical cord lesions, which, like skeletal injury, may coexist with CBP although no cases were reported in this survey. Other rare diagnoses to be considered are acquired brachial neuropathy due to sepsis, arthrogryposis, and tumours such as infantile myofibroma. Clearly careful clinical assessment and observation, as well as appropriate imaging, are required to exclude these alternative diagnoses, but the diagnosis of CBP is usually straightforward. This survey supports the view that, in the vast majority of cases, CBP is the result of intrapartum injury, although in an appreciable minority there was no identifiable indicator of the application of unusual force in the course of the delivery.

Upper plexus lesions (Narakas’s Groups I and II), representing between 42% and 87% of cases in other reports, were found in 91% of our cases. There were 6.3% complete plexus lesions (Narakas’s Groups III and IV). The higher proportion of more extensive lesions in previous reports may reflect a bias for the more extensive and severe lesions in studies originating from referral centres in contrast with this population based survey.

We recognise that this study may underestimate the incidence of CBP by up to 11% in view of the unreturned surveillance cards and questionnaires. It is disappointing that the incidence of this acquired condition remains so similar to that reported by Adler and Patterson in 1962.

The reported occurrence of full recovery in CBP varies widely from 13% to 80% of cases. Another study reported good functional recovery in 95% of infants, with 90% of those recovering showing improvement by 4 months of age. Our study found that at 6 months, 52% of infants with CBP had

Figure 2 Summary of outcomes.

<table>
<thead>
<tr>
<th>Movement</th>
<th>Cases</th>
<th>Full</th>
<th>Partial</th>
<th>None</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shoulder abduction (C5)</td>
<td>127</td>
<td>49</td>
<td>44</td>
<td>14</td>
</tr>
<tr>
<td>Elbow flexion (C6)</td>
<td>127</td>
<td>59</td>
<td>44</td>
<td>14</td>
</tr>
<tr>
<td>Elbow extension (C7)</td>
<td>125</td>
<td>59</td>
<td>44</td>
<td>14</td>
</tr>
</tbody>
</table>

Figure 3 Pattern of recovery.
recovered fully, but that there was no or partial recovery in 48%. Thus, in this population survey over a 12 month period, a large number of infants (133 out of 276) had considerable impairment at about 6 months of age. The range in age at assessment was 18–27 weeks (median 23) and is unlikely to have skewed the data, as we are unaware of any infants being excluded during this time interval. The prognosis for infants with extensive (Narakas’s Group III and IV) lesions was particularly poor. They are, however, a relatively small group, suggesting that it would be reasonable for them to be referred early for assessment at a specialised centre, together with those with slow recovery in early infancy. The assessment could include neurophysiological studies, the role of which in the evaluation of the extent and severity of CBP in the infant is currently uncertain. There have been some encouraging reports in recent years, however. For example, Smith reported that forearm electromyography and mixed nerve action potentials in 30 infants with CBP at a mean age of 4 months accurately predicted the nature of the lesion. In particular, she identified a group of infants with clinical signs of poor recovery, with conduction block indicating neuapraxia; they were managed conservatively with good outcomes but would have proceeded to surgery on clinical grounds alone. The establishment of specialist assessment and treatment centres in the United Kingdom could facilitate further evaluation of the role of neurophysiological studies.

There would seem to be two possible approaches to this clinical problem of CBP. One is to aim to reduce the number of infants with severe CBP. It would seem that fewer of these large and therefore vulnerable infants are being delivered by caesarean section than ought to be. The prediction that a fetus is going to be large should alert clinicians to the possibility that brachial palsy may be acquired in the course of delivery, and elective caesarean section should be considered. A second approach is to establish best practice when a baby is born with severe CBP. Data from this study suggest that this acquired condition will not disappear, and there continues to be uncertainty about the appropriate management of infants who are born with CBP and the place for specialist assessment and intervention by microsurgery. To be feasible, a randomised controlled trial of microsurgical nerve repair in infants with Group III or IV CBP with no recovery of function at the shoulder or elbow at 6 months of age should aim to achieve a reduction in the number who subsequently show no improvement (say from 32% to 5%); the demonstration of a smaller reduction would take too long given the relative rarity of the condition. Nevertheless, given the doubts that exist about the appropriate management of this debilitating condition, serious consideration should be given to a formal clinical trial.

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Contributors: G E-J, S P K and A M W designed the study. G E-J submitted the proposal for the survey to the British Paediatric Surveillance Unit. A W, G C, and A B are clinical academic assistants to S P K, Department of Plastic, Reconstructive and Hand Surgery, St James’s University Hospital, Leeds, and they circulated the questionnaires and managed the database. C H is a senior house officer in plastic surgery and assisted in the analysis of the follow up data. A M W and S P K both collaborated with G E-J in preparing the manuscript, with A M W providing the statistical analysis.

References


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