Differences in risk factors between early and late diagnosed developmental dysplasia of the hip

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ORIGINAL ARTICLE

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Correspondence to: Ms Sharpe, South Australian Birth Defects Register, Women's and Children's Hospital, 72 King William Road, North Adelaide, SA 5006, South Australia; cywhs.sabdr@cywhs.sa.gov.au

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Background: Developmental dysplasia of the hip (DDH) is common, affecting 7.3 per 1000 births in South Australia. Clinical screening programmes exist to identify the condition early to gain the maximum benefit from early treatment. Although these screening programmes are effective, there are still cases that are missed. Previous research has highlighted key risk factors in the development of DDH.
Objective: To compare the risk factors of cases of DDH identified late with those that were diagnosed early.
Methods: A total of 1281 children with DDH born in 1988–1996 were identified from the South Australian Birth Defects Register. Hospital records of those who had surgery for DDH within 5 years of life were examined for diagnosis details. Twenty seven (2.1%) had been diagnosed at or after 3 months of age and were considered the late DDH cases (a prevalence of 0.15 per 1000 live births). Various factors were compared with early diagnosed DDH cases.
Results: Female sex, vertex presentation, normal delivery, rural birth, and discharge from hospital less than 4 days after birth all significantly increased the risk of late diagnosis of DDH.
Conclusions: The results show differences in the risk factors for early and late diagnosed DDH. Some known risk factors for DDH are in fact protective for late diagnosis. These results highlight the need for broad newborn population screening and continued vigilance and training in screening programmes.

Developmental dysplasia of the hip (DDH) is a common congenital abnormality that affects the developing hip joint of the newborn. DDH refers to a spectrum of disease, including hips that are unstable, subluxated, dislocated, and/or have malformed acetabula.[1] Traditionally, radiological examination has been used to diagnose DDH; however, some hips that have acetabular dysplasia but are enlocated could be grouped with hips that are truly dislocated.

Over the last two decades, ultrasound has also been used to detect DDH. Ultrasound has the potential to identify minor abnormalities that are likely to resolve spontaneously without treatment. Therefore this method of diagnosis may lead to an apparent exaggeration of affected neonates.

Recent studies from the United Kingdom have shown that screening for DDH using ultrasound detected more cases than routine physical examination, but resulted in more children being treated.[2] A number of unfavourable treatment outcomes has also been shown from treatment of unaffected children with a false positive screening result.[3] Conversely, the results from the Medical Research Council's (MRC) United Kingdom hip trial showed that the use of ultrasound in infants with screen detected clinical hip instability was not associated with an increased risk of surgical treatment by 2 years of age.[4]

In South Australia, the Ortolani and Barlow tests are the basis of routine clinical examination screening programmes designed to detect DDH as soon after birth as possible.[5 6] These tests are performed at birth, on each day during the hospital stay, and at well baby clinics at 6 weeks and 3, 6, and 12 months of age. Ultrasound examination is only used if the clinical examinations in the first few months are equivocal. DDH cases in the South Australian Birth Defects Register include dislocated/dislocatable and subluxated/subluxatable hips, and the prevalence in the study period was 7.3 per 1000 live births. If mild degrees of acetabular dysplasia in stable hips are included, an earlier study in one year (1991) obtained a prevalence of 10.8 per 1000 births. Despite this rigorous screening, each year a few patients with DDH are not detected in the first 3 months of life and are considered late diagnosed. The late diagnosed DDH cases in this study were all true dislocations, with the femoral head dislocated outside the acetabulum, confirmed on ultrasound or radiographs.

There are several studies in the literature that report rates of late diagnosis of DDH.[7–14] Interpretation and comparison of rates of late diagnosed DDH is often difficult, especially in relation to the strict definition and age at diagnosis. Reported rates can range from 0.07 to 2.0 per 1000 births.[10] The cut off age for inclusion for late diagnosis is not consistent in the literature and can vary from 6 weeks to 20 months.[6 9] Palmen[14] reported a prevalence of late diagnosed DDH in the presence of a well established neonatal screening procedure of 0.53 per 1000 in the period 1973–1976. They reported that one quarter of the late diagnosed cases had not been diagnosed by 7 months of age and that 3% were diagnosed after 2 years. In a study from the United Kingdom in 1991–1995, late DDH cases were defined as cases presenting after the age of 6 weeks.[9]

Our earlier work in South Australia involving 916 DDH cases in 1988–1993 found that 55 cases required surgical treatment in the first 5 years of life.[13] Twenty two of these were late diagnosed at or after 3 months, and 33 were diagnosed before 3 months. If we assume that all late diagnosed cases required surgery, the proportion of early diagnosed cases that required surgical treatment was only 3.7% (33 out of 894). This shows the importance of early diagnosis.

A single study in Canada examined the differences in epidemiological characteristics between early and late diagnosed cases of DDH.[7] The authors did not report an overall population prevalence of DDH, but found that 21% had been diagnosed late, with the cut off point for late diagnosis being 20 months. Some significant epidemiological differences were found. Right sided DDH was significantly higher in the late diagnosed group ($p<0.0002$) as were cases of

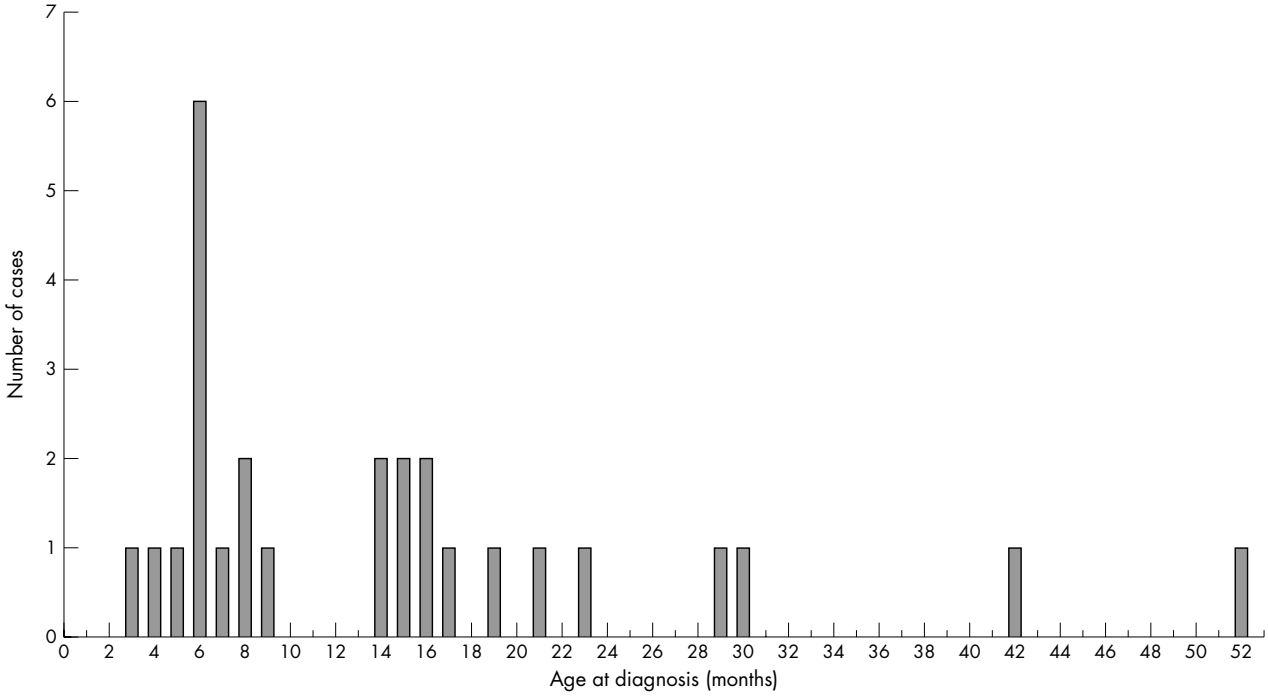

Figure 1 Age at diagnosis of developmental dysplasia of the hip.

bilateral DDH (p = 0.006). There were more female infants with DDH in the late diagnosed group, but the difference was not significant (p = 0.09).[7]

South Australian studies have previously confirmed breech presentation and female sex to be risk factors for DDH.[15 16] As there have been few studies on factors that increase the risk of late diagnosis, the aim of this study was to identify specific differences in the epidemiology of early and late diagnosed DDH.

MATERIALS AND METHODS

In South Australia, details of mother and baby have been routinely provided since 1981 on a perinatal data collection form by midwives to the Pregnancy Outcome Statistics Unit of the South Australian Department of Health. The data cover more than 99.9% of all births and include congenital abnormalities diagnosed at birth. Notifications of congenital abnormalities at birth have been supplemented by notifications up to the child's 5th birthday to the South Australian

Table 1 Crude odds ratios for various factors associated with late diagnosed developmental dysplasia of the hip

Variable	Cases (n = 27)	Controls (n = 1254)	Crude OR (95% CI)	p Value
Presentation				
Vertex	25	872	1.00	
Breech	1	369	0.09 (0.00 to 0.58)	0.008
Face	0	9	Undefined	
Unknown	1	4	–	
Method of delivery				
Spontaneous vaginal	18	537	1.00	
Other vaginal (including ventouse and forceps)	5	232	0.64 (0.18 to 1.83)	0.52
Caesarean section	3	481	0.19 (0.04 to 0.64)	0.005
Unknown	1	4	–	
Sex of baby				
Male	1	277	1.00	
Female	26	973	7.40 (1.20 to 304.54)	0.04
Unknown	0	4	–	
Hospital category				
Rural	10	173	3.89 (1.61 to 9.25)	0.002*
Home birth	1	0	Undefined	
Metropolitan	16	1077	1.00	
Unknown	0	4	–	
No of days before discharge from birth hospital (1 home birth excluded)				
<4 days	7	123	3.38 (1.17 to 8.58)	0.012*
⩾4 days	19	1127	1.00	
Median	4	6		

Data for one case and four controls were not complete.
*Fisher's exact test.
OR, Odds ratio; CI, confidence interval.

Birth Defects Register since 1986. These supplementary data are gathered from hospitals, special investigation and treatment centres, and practitioners treating children. These data are collected under specific legislation. Research conducted by the South Australian Birth Defects Register has been approved by the research ethics committee of the Women's and Children's Hospital, Adelaide, South Australia.

Notifications of DDH were retrieved from the South Australian Birth Defects Register for children born in 1988–1996, yielding 1281 cases of DDH from 176 427 live births. The State's database for inpatient separations (for 1988–2001) was used to identify all children born in 1988–1996 who had surgery for DDH within the first 5 years of life. This long follow up period of the Birth Defects Register restricted the number of years able to be included in the study to only those where five years of ascertainment had been completed—that is, up to 2001. Hospital medical records of the children who had surgery for DDH were reviewed to determine the timing and circumstances of diagnosis. In 27 cases, the diagnosis was made at or after 3 months of age, and these cases were considered late diagnosed DDH. They were all true dislocations with the femoral head dislocated outside the acetabulum, confirmed by radiographs. In some instances, DDH had been diagnosed early but treated late, or conservative treatment had been unsuccessful so surgery followed. These were not included in the late diagnosis group. The remaining 1254 DDH diagnoses were used as affected controls in the epidemiological analysis for potential risk factors. Cases and controls were linked to the South Australian Pregnancy Outcome Statistics Unit perinatal database by an identifier, common to both data sets. Maternal characteristics, pregnancy, and delivery details and complications were retrieved. Mother's race, mother's age, birth hospital type, previous births, baby's sex, presentation at delivery, method of delivery, birth weight, gestation, and length of hospital stay were compared.

Agreement between the perinatal data collection and hospital medical records has previously been established to be high, with κ values of 0.85 to 1.0 for risk factors examined in this study.[17]

RESULTS

In this series, 27 of 1281 cases of DDH in babies born between 1988 and 1996 were diagnosed at or after 3 months and were categorised as late diagnosed DDH. The prevalence of late diagnosed DDH over this nine year period was 0.15 per 1000 live births. In this group, there was a female preponderance (26:1). There were 21 unilateral cases of DDH; 18 were on the left and three on the right. There were six cases of bilateral DDH. The mean age at diagnosis was 14.2 months (range 3–52) (fig 1).

Table 2 Method of initial diagnosis of late diagnosed developmental dislocation of the hip

Method	No (%)
Family noticed limp when child started to walk:	12 (44)
Restriction in hip movement	4
Dragging leg	2
Waddling or limping gait	6
First noticed by child and youth health or family practitioner	9 (33)
Incidental finding during examination for other conditions	2 (7)
Unknown method of diagnosis	4 (15)
Total	27

Presentation at delivery

If cases in which presentation at delivery was not known are excluded, 25 out of 26 cases (96%) had a vertex presentation compared with 872 (69.8%) of the controls. There was one case (4%) with breech presentation in the late DDH group compared with 369 (29.5%) controls (odds ratio (OR) = 0.09 (95% confidence interval (CI) 0.0 to 0.58), p = 0.008), indicating that breech presentation is protective for late DDH (table 1), an interesting result considering that breech presentation is a well known risk factor for DDH.

Method of delivery

When compared with early diagnosed DDH, cases in which the baby was delivered by caesarean section had a significantly reduced crude odds ratio for late diagnosis (OR = 0.19 (95% CI 0.04 to 0.64), p = 0.005; table 1). Other vaginal deliveries were also at reduced risk (OR = 0.64 (95% CI 0.18 to 1.83)) for late diagnosis; however, this did not reach statistical significance.

Sex of baby

Twenty six (96%) of the 27 infants with late diagnosed DDH were female compared with 973 (77.8%) controls (OR = 7.40 (95% CI 1.20 to 304.54), p = 0.04; table 1). This highlights the added increased risk for female babies.

Hospital category

In 10 of the 27 late DDH cases (37%), birth took place in a rural hospital compared with only 173 (13.8%) of the controls (OR = 3.89 (95% CI 1.61 to 9.25), p = 0.002, table 1), indicating that late diagnosis of DDH was nearly four times more likely to occur in a rural hospital than a metropolitan hospital.

Length of stay

The median length of stay in the late DDH cases was four days compared with six days in the controls. Babies discharged within four days were nearly three and a half times (OR = 3.38) more likely to have late diagnosed than early diagnosed DDH. This finding was significant (p = 0.012; table 1).

Maternal Factors

Maternal factors such as age, race, marital status, and number of previous births, as well as baby's birth weight and gestation were compared between the cases and controls. No significant differences were found.

Method of initial diagnosis

In this series of 27 late diagnosed cases of DDH, most cases were initially identified by the parents and were then referred for medical examination and further investigation. Others were first noticed by a child and youth health professional or by the child's local general practitioner. There were four cases where the initial method of diagnosis was not known. Two others were incidental findings, detected during examination for other conditions (table 2).

DISCUSSION

The epidemiology of late diagnosed DDH has received little previous attention in the literature, and direct comparison between various studies is difficult for many reasons. Firstly, the cut off age for diagnosing late DDH can range from 6 weeks to 20 months.[6 8 13] The age at which a child presents with the condition will influence the ease of treatment, with early diagnosis and treatment usually ensuring a good result and preventing early hip osteoarthritis in adult life. Earlier studies have shown that early detection also reduces the need for surgical intervention.[13 18]

The prevalence of late diagnosed DDH in South Australia was much lower than other reported studies, with 0.15 cases per 1000 live births compared with 0.53 and 0.47 per 1000 live births in other studies.[8 14] This highlights the effectiveness of the South Australian clinical neonatal screening programme in detecting cases of DDH early. However, it is difficult to compare these figures directly as the definitions of late DDH are not consistent in the literature and we have only included cases of late diagnosis requiring surgery.

Babies with a breech presentation had a reduced risk of late diagnosis of DDH (OR = 0.09, p = 0.008). It is possible that these babies were more closely examined in the neonatal period for DDH, with the treating paediatrician/neonatologist knowing that breech presentation babies are at higher risk of DDH. In addition, there may be some referral of rural babies with breech presentation to larger metropolitan hospitals, resulting in closer neonatal examination.

Caesarean section deliveries also had a reduced risk of late diagnosis of DDH (OR = 0.19, p = 0.005). A baby is more likely to be examined by an experienced paediatrician/neonatologist after a caesarean section delivery than after a routine vaginal delivery. This may contribute to the ''protective'' effect of caesarean section.

Rural births had a four times increased risk of late diagnosed DDH compared with metropolitan births. The reason for this difference may be that rural practitioners, who deliver smaller numbers of babies per year than their metropolitan colleagues and have a high turnover rate, would have reduced opportunity to examine as many babies by the Barlow and Ortolani manoeuvres. This implies that training in clinical examination and familiarity with the use of that examination technique is of paramount importance. This may be especially relevant to areas where medical staff have lower exposures to neonates or areas where there may be higher staff turnover.

Female infants have been shown to be at even greater risk of late diagnosis than early diagnosis (OR = 7.4). This has been reported in other studies.[8] The reason for this repeated finding remains unknown. Some authors have implied a female susceptibility to relaxin hormone, and more recently a possible relaxin hormone receptor sensitivity has been queried.[19]

Infants discharged from hospital relatively early (< four days) after delivery were also found to be significantly more at risk of having late diagnosed DDH. With the increasing demand for hospital beds, earlier discharges to community based supports are favoured in many situations. Over the study period, the mean length of stay for all babies born at term (≥37 weeks) in South Australian hospitals decreased from 5.8 days to 4.3 days.[20] This reduction may inevitably result in a small group of babies that are not examined as often as those with longer hospital stays. A shorter hospital stay provides the clinician with fewer opportunities to examine a baby in a relaxed state (which is paramount for a proper Ortolani/Barlow clinical examination).

Whether late diagnosed DDH has the same aetiology as DDH diagnosed earlier remains unknown. However, significant epidemiological differences exist between the two groups. The findings that breech presentation and caesarean delivery are protective may be due to these babies receiving greater medical attention.

One limitation of this study is that the number of cases of late diagnosed DDH is small. However, significant differences have been identified, with female sex, rural birth, vertex presentation, vaginal delivery, and early discharge increasing the risk of late diagnosis. Some of the results of this study, such as that pertaining to rural births, may not be applicable to other populations. However, the emphasis of continued clinical examination training could be applied to any area with a high medical staff turnover. Clinical examination of a neonate's hips is a specific skill that requires a relaxed baby and an experienced clinician. Specific training for rural centres appears indicated in South Australia to minimise the late diagnosis rate.[21]

Clinicians appear to be achieving early diagnosis of DDH in cases of breech presentation and caesarean deliveries. They must, however, assume that all babies have DDH until proven otherwise, especially with our findings that normal, female, vaginal deliveries are most at risk of late diagnosis.

What is already known on this topic

- Female babies, breech presentation, primiparity, and family history are well known risk factors for early diagnosed DDH
- Late diagnosis of DDH often leads to surgical treatment and a poorer outcome

What this study adds

- It shows how a centralised healthcare system with clinical population screening can reduce the rate of late diagnosed DDH to 0.15 per 1000 live births
- The risk factors for late diagnosed DDH are highlighted and shown to be different from those associated with early diagnosed DDH

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Authors' affiliations

P Sharpe, South Australian Birth Defects Register, Women's & Children's Hospital, Adelaide, Australia
K Mulpuri, P J Cundy, Department of Orthopaedic Surgery, Women's and Children's Hospital, Adelaide
A Chan, Pregnancy Outcome Statistics Unit, Epidemiology Branch, Department of Health, Adelaide

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