

Selective sonographic screening for developmental dysplasia of the hip – increasing trends in late diagnosis

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There are concerns that selective sonographic screening for developmental dysplasia of the hip (DDH) may be suboptimal. Our aim was to test this hypothesis by identifying trends in presentation and surgical treatment in patients with DDH. This is a retrospective review of children born between 1997-2018 who were treated surgically for DDH at our sub-regional paediatric orthopaedic unit. Demographic data, risk factors, age of diagnosis and surgical treatments were analysed. Late diagnosis was defined as greater than 4 months. 103 children (14 male, 89 female) underwent surgery. 93 hips were operated for dislocation and 21 for dysplasia. 13 patients presented with bilateral hip dislocations. The median age at diagnosis was 10 months (95% CI: 4-15). 62/103 (60.2%) were diagnosed late (after 4 months) and the median age for diagnosis in this group was 18.5 months (95% CI: 16-20.5). Significantly more patients were referred late ($p=0.0077$). The presence of risk factors (breach presentation or family history) was associated with early diagnosis. Over the duration of our study the operation rate per 1000 live births gradually increased, and on Poisson regression analysis there was a statistically significant increasing trend towards late diagnosis in recent years ($p=0.0237$), which necessitated more aggressive surgical management. In the UK, the current selective sonographic screening programme for DDH has shown a deterioration over the years of this study and this questions its current effectiveness. It appears that the majority of irreducible hip dislocations are diagnosed late, with an increased need for surgical management.

Keywords : Developmental Dysplasia of the Hip, DDH, surgical management of DDH, DDH hip screening.

INTRODUCTION

Developmental dysplasia of the hip (DDH) is an important orthopaedic condition that mainly presents in newborns and infants. DDH embodies a spectrum of diseases ranging from hip dysplasia to irreducible hip dislocation.

The incidence of DDH varies between racial and geographic locations¹. A recent UK study of 37,233 patients quotes the incidence of pathological DDH as 4.9 per 1,000 live births². There are multiple risk factors for DDH, the most important two being positive family history and breach presentation³. The condition is more common in females and on the left side^{1,2,4}.

The initial diagnosis of pathological DDH in the neonate and infant includes clinical examination (positive Ortolani and Barlow manoeuvres, or unilateral limitation of hip abduction), ultrasonographic imaging and/or radiographs of the hip joint. Early detection (before 4 months of age)

may lead to successful treatment with less invasive modalities, such as bracing with a Pavlik harness. Late diagnosis (after 4 months of age) may require operative intervention, such as closed or open hip joint reduction (plus hip spica immobilisation). More complex surgical procedures such as femoral and pelvic osteotomies may be necessary in children over the age of 18 months. Orthopaedic problems in adolescents and adults from long term untreated irreducible hip dislocation, include severe hip dysplasia, back pain or premature osteoarthritis of the hip joint⁵⁻⁷. One third of total hip replacement surgery under the age of 60 years of age is thought to be secondary to hip dysplasia⁸.

In the United Kingdom (UK), the NHS Newborn and Infant Physical Examination (NIPE) programme has guidelines on selective sonographic screening for pathological DDH⁹. There are 3 major screening recommendations:

1. All newborns are screened within 72 hours of birth with the Ortolani and Barlow manoeuvres. ‘Screen

positive' results necessitate ultrasonographic assessment within two weeks.

2. NIPE guidelines also advise that 'high risk' (first degree family history or breech) babies should undergo sonographic assessment between 4 to 6 weeks.
3. 'Screen positive' hip instability manoeuvres by the General Practitioner at the 6 to 8 week infant examination should be referred directly to paediatric orthopaedic surgeon for urgent expert opinion and be seen by 10 weeks of age.

The effectiveness of the NIPE DDH hip screening programme has been questioned and there are concerns that it may be suboptimal in meeting the World Health Organisation's screening criteria¹⁰⁻¹⁴. We therefore decided to test this hypothesis by identifying trends in presentation, diagnosis and surgical treatment of patients with DDH in our sub-regional paediatric orthopaedic centre.

MATERIALS AND METHODS

This is a retrospective review of 103 children who were operated upon for pathological DDH in a sub-regional paediatric orthopaedic unit, serving a population of 530,000 and in which approximately 25% of the population are under the age of 16 years¹⁵. Data was collected from hospital electronic, radiological and written records. Inclusion criteria included all cases of pathological DDH (dysplasia or dislocation) that required surgical intervention (closed or open hip joint surgery). All patients were from our hospital's catchment area, with no refugee or other fragility groups.

The sources of referral of cases of pathological DDH requiring surgery were from GPs, paediatricians, and one case through the Emergency Department.

Due to cases of delayed presentation information was analysed from the date of birth not at the age of diagnosis. Patients included in this study were born between 1st January 1997 and 31st December 2018. The authors defined early presentation/diagnosis as within 4 months of birth^{3,16}.

To assess the operation rate per live births in our hospital's catchment area, we collected birth rate data from the UK Government's Office for National Statistics (ONS) website¹⁷.

GraphPad Prism 8.4.2 was used for the statistical analysis. Normality was assessed using the Shapiro-Wilk test. Comparison between sample groups was conducted using one-way ANOVA (Kruskal-Wallis with multiple comparisons) and the Mann-Whitney

U-test. To compare numbers of early and late referrals each year we used the Wilcoxon matched-pairs signed rank test. To examine the relationship between early and late diagnosis, Poisson regression analysis was performed. Statistical significance was set at $p < 0.05$.

RESULTS

103 children (14 male, 89 female) underwent surgical intervention for DDH. 21 presented with pathological dysplasia and 82 presented with dislocation. Bilateral hip dislocations were diagnosed in 13 patients. Two patients with bilateral dislocations had one dislocated hip joint successfully treated in Pavlik harness. In total 114 hips required surgery, 93 for dislocation and 21 for pathological hip dysplasia.

The median age of diagnosis in this study was 10 months (95% CI: 4-15). 41 patients (39.8%) were referred and diagnosed early (less than four months of age) and 62 (60.2%) were diagnosed late (after four months of age). The median age of diagnosis in the late diagnosis group was 18.5 months (95% CI: 16-20.5). When comparing early and late referrals each year, we found that significantly more patients were referred late ($p = 0.0077$).

93 dislocated hips were treated surgically (see Table I). 25 were treated with closed reduction only, at a median age of 5 months (95% CI: 4-10.5). 34 were treated with open reduction only, at a median age of 11 months (95% CI: 10.8-14). 34 hips underwent primary open reduction with pelvic and/or femoral osteotomy, at a median age of 23 months (95% CI: 21-25).

Of the 25 dislocated hips treated with closed reduction only, the median age at diagnosis was 2.4 months (95% CI: 0.5-4). Of the 34 hips treated with open reduction only, the median age at diagnosis was 4.5 months (95% CI: 2.5-12). Of the 34 hips treated with pelvic and/or femoral osteotomy, the median age at diagnosis was 20 months (95% CI: 18-22). On ANOVA testing, the age at diagnosis was significantly different for each of these surgical treatments ($p < 0.0001$). This shows that a delay to diagnosis of dislocation was associated with a need for more aggressive surgical intervention.

16 patients treated with closed or open reduction subsequently required a secondary pelvic and/or femoral osteotomy, at median age of 25 months (95% CI: 20.5-34). Four patients treated initially with closed reduction required revision to open reduction, at a median age of 10.5 months (95% CI: 10-12). Three patients initially treated with pelvic osteotomy

Table I. — Primary surgical procedures for pathological DDH.

	Number	Median age of diagnosis, months (95% CI)	Median age of surgery, months (95% CI)
Dislocated hips			
Closed Reduction	25	2.4 (0.5-4)	5 (4-10.5)
Open Reduction	34	4.5 (2.5-12)	11 (10.8-14)
Pelvic/Femoral Osteotomy	34	20 (18-22)	23 (21-25)
Dysplastic hips			
Closed Reduction	12	2.5 (1-9)	10 (6-13)
Open Reduction	1	7	8
Pelvic/Femoral Osteotomy	8	28 (15-45.5)	40 (21-63)

required a secondary femoral osteotomy, at a median of 26 months (95% CI: 22-27)

21 dysplastic hips were treated surgically (see Table I). 12 were treated with closed reduction, at a median age of 10 months (95% CI: 6-13). One hip was treated with open reduction at 8 months. Eight hips underwent primary open reduction with pelvic and/or femoral osteotomy at a median age of 40 months (95% CI: 21-63).

Of the 12 dysplastic hips treated with closed reduction only, the median age at diagnosis was 2.5 months (95% CI: 1-9). The one hip treated with open reduction was diagnosed at 7 months. Of the 8 hips treated with pelvic and/or femoral osteotomy, the median age of diagnosis was 28 months (95% CI: 15-45.5). On ANOVA testing, the age at diagnosis was significantly different for each of these surgical treatments ($p=0.0047$). This shows that a delay to diagnosis of dysplasia was associated with a need for more aggressive surgical intervention.

One patient treated initially with closed reduction subsequently required a secondary pelvic osteotomy at 27 months.

In our cohort of 103 patients managed surgically for DDH, 10 (9.7%) had a family history of DDH and 9 (8.7%) were breech presentation. Two patients had both positive family history and were breech presentation. The median age at diagnosis of those with no risk factors was 13 months (95% CI: 11-17). The median age at diagnosis of those with a positive family history was 2 months (95% CI: 1.3-7.7), which was significantly less than those with no risk factors ($p=0.0454$). The median age at diagnosis of those with breech presentation was 1.9 months (95% CI: 1.1-2.7), which was significantly less than those with no risk factors ($p=0.0164$).

Over the 22 years of our study there was a gradual increase in trend in the number of operations per 1000 live births, from 0.74 to 0.91 (see figure 1). On Poisson regression analysis, looking at the relationship between early and late diagnosis from the years 1997 to 2018, there was a statistically significant trend towards late diagnosis in recent years ($p=0.0237$; see figure 2).

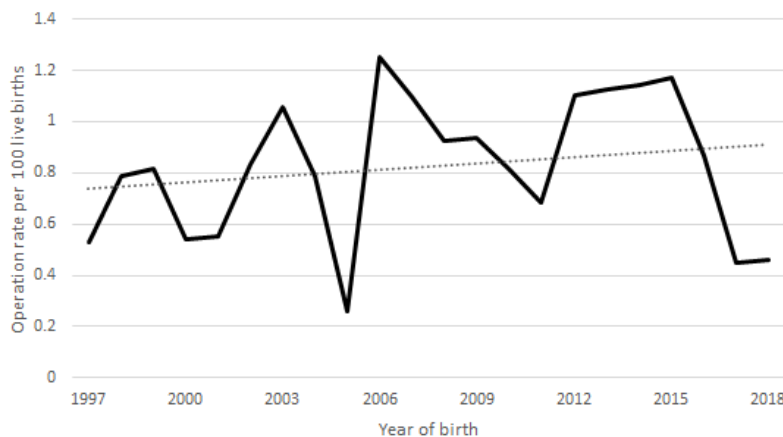


Fig. 1 — Operation rate per 1000 live births, showing a gradual increasing trend over the 22 years of our study.

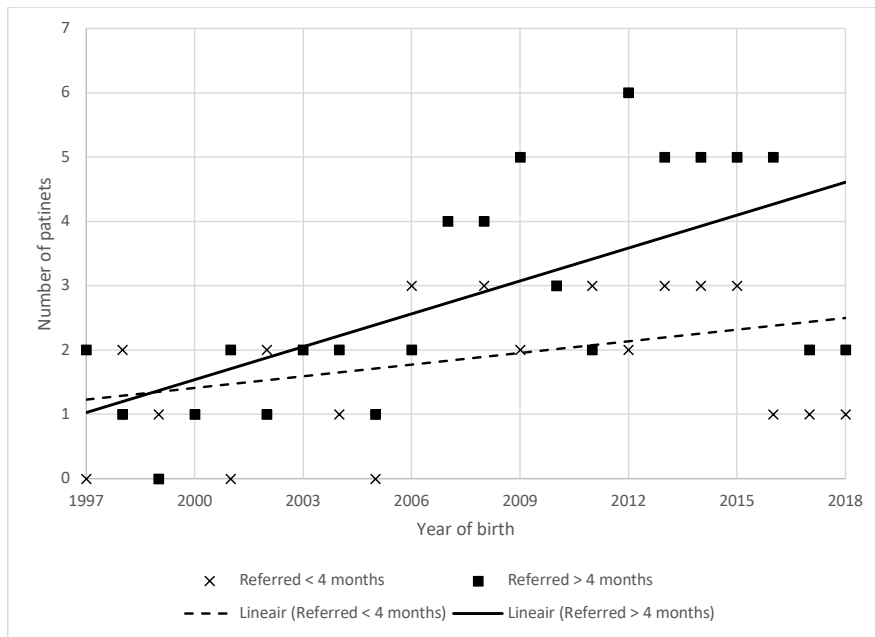


Fig. 2 — Number of early (<4 months) and late (>4 months) diagnoses per year. Note the increasing trends towards late referral in recent years.

DISCUSSION

The results of our study show that from 1997 to 2018, for patients managed surgically for pathological DDH (dysplasia and dislocation), there has been an increasing statistically significant number of those presenting and being diagnosed late (compared to the earlier years of the study). This delay to diagnosis for patients with both dysplasia and dislocations has been matched with an increasing operation rate per 1000 live births, and also necessitated increasingly aggressive surgical management.

There is no international consensus on the optimal screening programme for the diagnosis of DDH. Debate exists on which technique is superior: clinical examination alone, clinical/selective ‘at risk’ sonographic assessment, or universal sonographic hip imaging. At present there is an insufficient research evidence base to support one method over another statistically^{12,13,18,19}. The AAOS systematic review noted that although there were almost 4000 research papers on the subject of DDH screening, over 95% of studies had significant weaknesses in design and methodology, making it difficult or impossible to draw conclusions on the optimum DDH hip screening method¹³.

Universal ultrasound screening may allow for early detection of DDH and may reduce the need for more aggressive surgical treatment^{20,21}. However, this method has been associated with a higher overall treatment rate¹⁸. Up to 7% of the population may be treated in

some studies²². In the UK, NIPE guidance largely relies on the universal clinical hip screening technique to identifying ‘screen positive’ hips, but this assumes a level of competence in detecting rare, subtle clinical features of instability that could easily be missed by somebody not wholly familiar with DDH^{10,23}. This screening method does not identify irreducible hip dislocation or significant hip dysplasia. The sensitivity and positive predictive values are relatively low and this universal clinical method cannot be regarded as an effective screening programme but as a sub optimal surveillance method due to its low accuracy^{3,23}.

We identified that children with risks factors for DDH (first degree family history or breech presentation) were, on average, referred and diagnosed earlier than those without risk factors; and as such this aspect of the selective screening programme must be commended. Ömeroğlu et al. identified that family history of DDH and/or breech presentation are associated with the development of sonographically more severe hip dysplasia, which reinforces the utility of identifying these key risk factors as part of a selective screening programme²⁴.

Our study is limited in that we have only considered late diagnosis in a cohort of surgically managed patients. Broadhurst et al. (2019) looked at late diagnosis in all DDH patients, noting an incidence of 1.28 per 1,000 live births (diagnosed > 1 year of age)¹⁴. This has remained largely unchanged since 1994 and, despite NIPE’s recommendations, late diagnosis of DDH in the UK remains problematic. In our study late

presentation was defined as after four months of age, to take into account screening at birth, the 6 to 8 week check by the General Practitioner and also to allow for delays with processing the referral and initial review in the Paediatric Orthopaedic clinic. There has been a progressive increase in the proportion of children that are referred late in our sub-regional DDH service despite an established selective screening programme based on the NIPE guidelines. This would suggest that the NIPE programme of screening to achieve early diagnosis is becoming less effective with time.

CONCLUSION

This study has identified an increasing trend in the late diagnosis of DDH, and also the operation rate per 1000 live births. This questions the effectiveness of the current NIPE selective sonographic screening programme in the diagnosis of DDH. Late diagnosis was associated with a need for more aggressive surgical treatment, such as open reduction and pelvic/femoral osteotomies.

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