Why is congenital dislocation of the hip still missed?

Analysis of 96,891 infants screened in Malmö 1956-1987

Jan Sanfridson[1], Inga Redlund-Johnell[2] and Alf Udén[1]

During 1956 through 1987, 96,891 children have been screened for neonatal hip instability according to the tests of Ortolani and Barlow. In 1956 through 1972 only 4/58, 759 (0.07 permille) were missed, whereas during 1980 through 1987, 12/19, 398 (0. 6 permille) were missed. This increase is not caused by any formal alteration of the screening programme. The screening has prevented a late diagnosis in all children born in breech presentation and in all boys except one. General factors such as female sex and joint laxity imply an increased risk for being missed in the screening, whereas mechanical factors such as breech presentation and the primogeniture effect likely facilitate an early diagnosis in the screening. The time between birth and the first examination is also of some importance.

Lund University, Departments of [1]Orthopedics and [2]Diagnostic Radiology, Malmö General Hospital, S-214 01 Malmö, Sweden. Correspondence: Dr. A. Udén
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Screening of newborns for hip instability and treatment with the von Rosen splint was introduced in Malmö in 1956. The excellent result of this program from the start until 1972 is well documented (Fredensborg 1976a). However, during the 1980's more infants have escaped an early diagnosis. We have analysed the routines of the screening and characteristics of the children who have presented with a late diagnosed congenital dislocation of the hip (CDH). The aim of the study was to identify risk groups and to improve our screening routines.

Patients and methods

During 1956 through 1987, 96, 891 children were born in Malmö. The number treated in a von Rosen splint because of neonatal hip instability (NHI) has varied over the years (Figure 1).

Neonatal screening

All newborns are examined at the maternity ward by a pediatrician around 9–10 am. During 1956 through 1963 the examination was performed twice a week and the children were examined only once. Since 1963 the examination was performed every day except on Sundays and holidays. Thus, the majority of children were examined within 24 hours, and they were examined once more on the day of discharge from the maternity ward. Until the middle of 1963 the Ortolani sign was used as criterion for CDH and thereafter the Barlow test was also used (Palmén 1961). All children with suspected instability were referred to the orthopedic department; if the instability could be confirmed, the child was treated in a von Rosen (1961) splint.

The maternity ward records of the children with a late diagnosed CDH have been analysed with respect to the experience of the examiner, the time between birth and examination, the findings noted, the type of presentation and birth-weight.

One hundred maternity ward records of children born in 1980 through 1987 were randomily chosen for comparison. Two of these children were excluded because they were treated with a von Rosen splint, leaving 98 non-treated newborns in the comparison group.

The radiographs of the patients with late discovered CDH have been reexamined. The examination had been performed with two anteroposterior films, one with 45° abduction, flexion and outward rotation of the femora and the second one with extended legs and inward rotation (Andrén 1962). The acetabular index was measured according to Lusted and Keats (1972). The dislocation has been evaluated as a difference in distance from the femoral head or the most proximal part of the femur to the symphyseal Y-line (Lusted and Keats 1972) and as a difference in distance from the cotyloid notch of the acetabulum to the centre of the femoral physis. The greatest diameter of the ossified caput was measured as well.

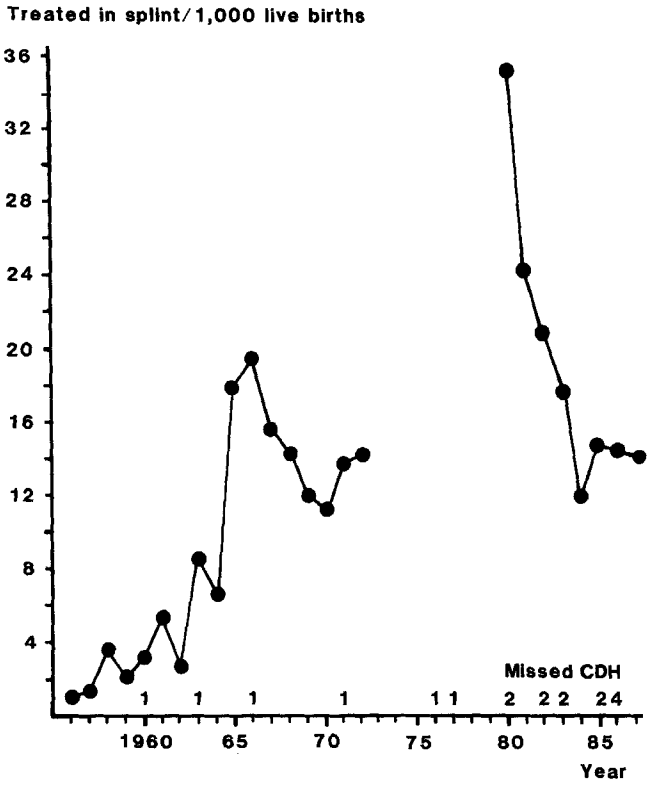

Figure 1. Number of cases per 1,000 newborns with NHI who were treated with a von Rosen splint. In 1963 the Barlow test was intro duced in the screening. The number above the bottom line denotes children born that year with late diagnosed CDH.

Table 1. Radiographic findings in infants with late diagnosed CDH. The order is chronological. Case 16 is the only boy

Case	Year born	Affected hip	Age at diagnosis (months)	Acetabular index[a]		Distance from symphyseal Y-line to caput femoris[b] (mm)		Distance between cotyloid notch and femur in Lauenstein (mm)	
				Right	Left	Right	Left	Right	Left
1	1960	dx	18	36*	22	+2	–6	21	15
2	1963	sin	18	20	40*	–7	+5	15	22
3	1966	dx+sin	1	26	26	–2	+1	14	13
4	1971	sin	14	23	52*	–9	+8	18	25
5	1976	sin	2	28	44*	–11	–8	15	18
6	1977	sin	36	25	32*	–7	+3	15	25
7	1980	sin	3	28	41*	–9	–5	14	16
8	1980	dx	2	38	30	–4	–10	16	13
9	1982	dx	14	28	15	+2	–6	25	13
10	1982	dx	15	32*	19	–2	–11	20	25
11	1983	dx	3	43*	31	–7	–9	17	14
12	1983	sin	3	32	45*	–8	–5	14	14
13	1985	dx	3	37*	29	–5	–10	film	missing
14	1985	sin	7	28	40*	–10	–6	14	16
15	1986	sin	2	25	30	–10	–9	15	18
16	1986	dx	2	33*	29	–9	–11	17	17
17	1986	dx+sin	5	36*	38*	–8	–7	17	17
18	1986	sin	4	25	37*	–11	–6	17	17

[a]The asterisk denotes an acetabular index above 2 SD (Tönnis 1976).
[b]Positive value above respectively negative value below that line.

Criteria for radiographic diagnosis of dislocation have been a difference in the measured distances of more than 2 mm and a difference in acetabular angle of more than 3° when unilateral, and according to visual impression of Shenton's line and of the femoral head when bilateral.

The parents or the patient herself were interviewed after having received a questionnaire. Joint laxity according to Carter and Wilkinson (1964) was recorded in the ten oldest patients at a reexamination during 1988. The mean age was 9.8 years (2–25).

Statistical tests used were Student's *t*-test for difference between means and Fisher's exact test.

Results

In all, 18 children have presented with late diagnosed CDH (0.2 permille). Since 1980 there is an apparent increase of the incidence (Figure 1). Among 19,398 born in 1980 through 1987, 12 were missed (0.6 permille) as compared with 0.07 permille during 1956 through 1972 (4/58, 759) ($P < 0.001$). The age at the diagnosis was lower in the 1980's, on the average less than six months, whereas the six first patients with a late diagnosis on the average were more than 14 months of age (Table 1). Of the 18 children with late diagnosed CDH only two had bilateral dislocation. In seven children there was a unilateral dislocation on the right side and in nine on the left side.

None of the children with late-diagnosed CDH was born in breech presentation, 7 were first-borns. Crepitation and/or a slight sliding was noted at the neonatal screening in 7 of the 18 patients. In the comparison group similar observations were made in 9 children. Thus, such findings were more frequent in babies with late CDH ($P < 0.001$). Six children with late CDH were born on a Saturday or on a day before a holiday which was twice as frequent as expected (NS). The time between delivery and the first examination was 22 hours (SD 13) in cases with a late diagnosis and 16 hours (SD 11) in the comparison group ($P < 0.05$).

During the years 1980 through 1987 the number of pediatricians who referred the children to the orthopedic department varied between 14 and 20. The most experienced pediatrician had examined 3 out of 12 children with a late diagnosis within 24 hours and checked 4 others within 5 days without noting any instability. None with a late diagnosis was examined by an unexperienced examiner only. Of the 18 children with a late CDH one was suspected by the pediatrician to have an unstable hip but since this could not be confirmed by the orthopedist she was not treated.

The mean birth-weight of the girls was 3,440 (2,600–4,700) g and the birth-weight of the boy was 2,520 g. During the first year of life 14 of the children usually lay prone in bed, three lay in a supine position and one in varying positions.

There was no strong familial occurrence of CDH. None of the parents had had CDH or NHI. One younger and one older sister of 17 siblings had NHI. One grandfather and one great-grandmother had CDH. In one case the sister of the father and her children had CDH.

The radiographic findings are presented in Table 1. The quotient right/left of the ala ossis ilii varied between 0.88–1.10. Thus, the rotation of the pelvis was so small that the acetabular index can be reliably measured (Tönnis 1976). In most cases there was a severe dysplasia and in all cases the hip was shown to be dislocated at least in one of the radiographs. When visible, the ossification center of the caput femoris was always smaller on the dislocated side.

In three of the patients aged 8, 11 and 25 years at the follow-up there was no increased joint laxity whereas in the other 7 at least 3 of the 5 joints tested showed an increased mobility.

Discussion

One explanation for the increase of late diagnosed CDH could be overdiagnosis. In the nation-wide Swedish material 1973–1979 of 407 cases with a late diagnosis three fourths had subluxation or dysplasia of the hip only (Palmén 1984), and similar proportions were reported in a Norwegian population (Bjerkreim and Johansen 1987). However, neither the clinical nor the radiographic criteria for CDH have changed during this study and all cases in our study had dislocated hips. Still, during the 1980's the children were younger at diagnosis so there is a small possibility that some of them would have developed normal hips without any treatment, in the same way as the dysplastic hips (Pratt et al. 1982). However, a real increase in late diagnosed CDH has definitely occurred.

All children with late diagnosed CDH were as newborns examined by an experienced pediatrician in spite of the fact that 14–20 different examiners/year were involved during the years 1980–87. The one with the most experience was not able to detect

a large proportion of the children with a late diagnosis. Thus, some cases of CDH were not possible to detect although the hips were checked by an experienced examiner in proper time. The time of examination seemed to be of some importance since children born on a Saturday or a day before a holiday ran a higher risk of not being diagnosed, and the children with late CDH were examined somewhat later than the other children. Crepitation or sliding was more frequent in newborns with late diagnosed CDH, so if they had been more thoroughly checked the diagnosis could perhaps have been made earlier. However, since similar remarks were made in 9 percent of the control group this would imply that 250 newborns per year in Malmö should be examined more thoroughly to avoid a late diagnosis in 0–1 child.

The more extensive examination introduced in 1963 with the Barlow test and an extra examination has not reduced the number of late CDH but rather increased the number of newborns treated with a splint. Maybe stretching of the joint capsule because of too much provocation can be a cause of CDH. Especially the second part of the Barlow test when the examiners try to push the femoral head out of the acetabulum could be harmful if done too harshly (Moore 1989). However, there is no evidence that the provocation would have been more harsh during the 80's.

There was no premature among those with late CDH. Thus, the increased risk of prematures in Gothenburg to escape an early diagnosis could be due to local screening routines (Hansson 1980).

In Malmö 32 percent of the children with NIH were born in breech presentation as compared with 6 percent in the general population (Fredensborg 1975). Interestingly, none of the 18 with late CDH in this study was born in breech presentation. A similar observation has been made in Bristol where all ten patients with a late diagnosis, in spite of screening, presented by vertex at birth (Dunn et al. 1985). Before screening was introduced, children born in breech presentation contributed to almost half of the children with CDH (Dunn 1976). Thus, it is obvious that children in breech presentation have an instability that is easier to detect during screening and it is possible that an adequate screen ing could prevent a late diagnosis in practically all such infants. In the national study of CDH in Sweden the frequency of breech presentation was low in cases with late diagnosis after screening was introduced (Palmén 1984) and the same observation has been made in Australia (Bower et al. 1987).

Before the era of screening 33 percent had bilateral CDH (Wynne-Davies 1970). In our screened population bilateral affection was rare in cases with late diagnosed CDH whereas 60 percent of newborns treated for NHI had bilateral affection (Fredensborg 1976b). A similar observation has also been made in Australia (Bower et al. 1987). One explanation could be that those with bilateral hip instability has a double chance to be detected at the screening since, as shown by sonography, hip dislocation can be diagnosed by sonography without being clinically demonstrable (Dahlström 1989). Another explanation could be a spontaneous normalization of one side (Pratt et al. 1982).

The joint laxity was markedly increased in late diagnosed cases. Even if there was no strict control group, compared with 110 healthy children with a mean age of 10. 3 years (Fredensborg 1975) joint laxity was more common in the wrist ($P = 0.001$), elbow ($P = 0.0003$), knee ($P = 0.01$) and in the ankle ($P = 0.00003$). Despite the limited value of a control group examined by another examiner the findings contradict the suggestion that increased joint laxity should just be connected with NHI and not with late diagnosed CDH (Wynne-Davies 1970).

In this series firstborns were equally or slightly less common than in the general population.

Although extra attention should be paid to children with heredity for CDH or NHI this was less important in our group, since only one of 53 first degree relatives had been treated for NHI at the time of birth of the proband. In Gothenburg 5/20 had heredity for CDH in their families (Hansson 1988).

One explanation for the female dominance in those with late CDH could be an altered connective tissue or an effect of hormones, since some girls have a more pronounced instability on the fifth day of life than on the first day, a phenomenon never observed in boys (Dahlström 1989).

Girls of normal birth-weight born in vertex presentation are still a problem with regard to late diagnosed CDH. An alteration of the screening programme could be one way to solve the problem, i. e. to examine all children only once during the first 24 hours of life in a more gentle way (Moore 1989) and accept a few cases with a late diagnosis.

Sonography has been used in the screening but a new program must be going on for several years before you can prove that it is superior to a clinical screening, especially compared to the early years of screening in Malmö.

The deterioration during the 1980's implies that the frequency of late diagnosed CDH in Malmö nowadays is in level with the rest of Sweden (Palmén 1984).

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