Screening for Developmental Dysplasia of the Hip: From Theory to Practice

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ABSTRACT. *Objective.* **To evaluate an organizational model for neonatal population screening for developmental dysplasia of the hip.**

Methods. **In 4648 neonates born in six hospitals of the Lombardy region, screening for developmental dysplasia of the hip was done using the Ortolani-Barlow maneuver and ultrasonography.**

Results. **The frequency of positive results of clinical and ultrasound examinations carried out in the hospitals varied considerably as a result of difficulties in the Ortolani-Barlow test reproducibility and in the low sensitivity of the clinical examination when compared to ultrasonography. Neonatal screening results implied a large number of subjects with a IIa hip, according to Graf's system; as these subjects require follow-up, the cost of this type of screening is high. Ultrasound findings were normal at 69 days of life in 88% and 75% of subjects, respectively, with unilateral and bilateral type IIa hip.**

Conclusion. **This study evaluated various organizational models for screening (for different time periods and for selected populations) in relation to the cost-benefit ratio and demonstrated the different problems that still impede identification of a correct screening model.** *Pediatrics* **1997;99(2). URL: http://www.pediatrics.org/cgi/content/full/99/2/e5;** ***congenital dislocation of the hip, developmental dysplasia of the hip, clinical screening, newborn, reliability, ultrasonography.***

ABBREVIATIONS. DDH, developmental dysplasia of the hip; RR, relative risk; CI, confidence interval.

Screening is frequently used for developmental dysplasia of the hip (DDH) as there is a preclinical period when diagnosis is possible. An appropriate therapeutic intervention during this period[1–7] can change the natural history of the disorder positively.

In the early 1980s, Graf[8,9] introduced the diagnosis of DDH diagnosis by ultrasonography. It avoids radiology, an invasive procedure that is not sensitive in the diagnosis of DDH in the neonate. In addition, ultrasonography reveals characteristics undetectable with the Ortolani-Barlow maneuver or radiography.

Most ultrasound screening studies have been population studies carried out on samples selected on the basis of risk factors and/or clinical examinations.[10–12] The rare studies on unselected populations have involved insufficient numbers of cases to assess the validity and accuracy of sonographic screening definitively.[13–16] The objective of our study was to evaluate an organizational model for neonatal population screening.

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Received for publication Oct 12, 1995; accepted Sep 16, 1996.

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METHODS

All subjects born between December 1989 and November 30, 1990 at six hospitals of the Lombardy region where routine ultrasonography DDH screening was available were considered for inclusion in the study. Excluded were those with multiple malformations or syndromes, those requiring intensive therapy, and those transferred to other hospitals. Two doctors from each hospital (expert doctors) took preliminary 5-day training course to improve their performance of the Ortolani[17]-Barlow[18] maneuver. All other doctors participated in a DDH training seminar with practical exercises using a dummy. For each baby, the following data were obtained by interview: gender, twin birth, parity, oligohydramnios, breech position at the third trimester of pregnancy, breech presentation, family history of DDH, and associated malformations (twisted foot, talipes calcaneovalgus due to position or structural defect, torticollis).

Information about DDH and the routine diagnostic program (Ortolani-Barlow maneuver and ultrasonography) were recorded on the recovery card.

During the first 4 or 5 days of life, each baby underwent the Ortolani-Barlow maneuver two or three times in the hospital. This was almost always done by different doctors, each of whom was blinded with respect to the results obtained by the other(s). The doctor who did the last Ortolani-Barlow maneuver recorded the result on the clinical record form. The maneuver was repeated by an expert doctor when positive results were obtained at one or more maneuvers.

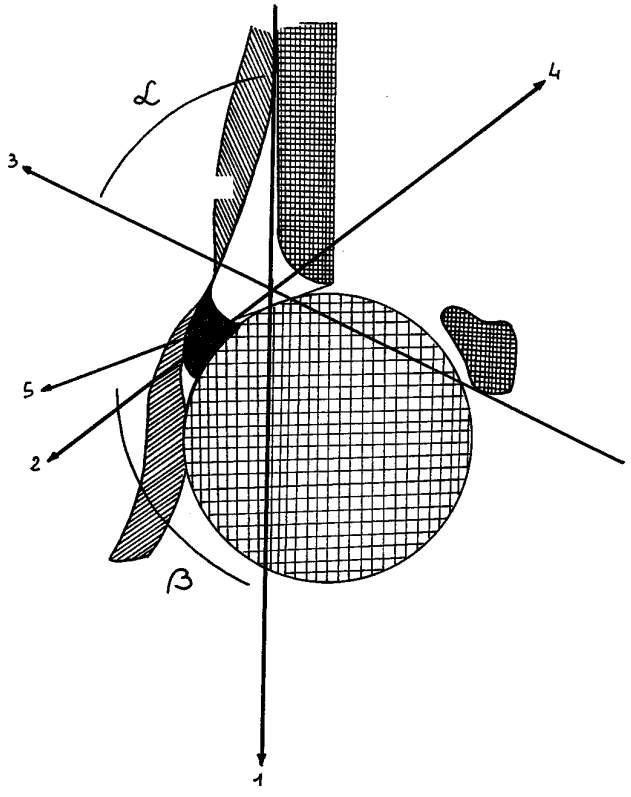
All subjects underwent sonography. An ultrasound screening examination using linear 5 and 7.5 Mhz probes was performed on all the babies within the first month of life. The examination was done by a doctor blinded to both the Ortolani-Barlow maneuver results and to any risk factors present. The doctors involved in DDH ultrasound examination had a mean of 27 months experience with it before the beginning of this study. The results were recorded according to Graf's system (Fig 1). Angles α and β were measured only when an anomaly was found (type IIa or worse). Sonography was repeated monthly on all babies with one or two type IIa hip(s) at the initial screening until their condition was found normal or appropriate therapy was decided.

The relation between clinical and ultrasonographic results was analyzed. Clinical agreement was analyzed by using the κ statistic; the accuracy (sensitivity, specificity, and predictive value) and the relative risk for the subjects with a positive Ortolani-Barlow maneuver were calculated. Data were stored and analyzed using a specific DB3 plus computer program.

RESULTS

Screened Population

A total of 4648 neonates, 2312 female (49.7%) and 2336 male (50.3%), were recruited at the six hospitals



Angle α	Angle β	Ultrasound Category
> 60°	< 55°	Ia
> 60°	> 55°	Ib
50° - 59°	> 55°	IIa (age < 3 mo)
50° - 59°	> 55°	IIb (age > 3 mo)
43° - 49°	< 77°	IIc
43° - 49°	> 77°	IId
< 43°	> 77°	III
< 43°	> 77°	IV

Figure. Angles according to Graf classification.

participating in the study. The mean gestational age was 40 weeks (range 27–42), the mean neonatal weight was 3260 grams (range 730–4850), and the mean maternal age was 29 years (range 15–53). Significant differences were not observed between the populations recruited from the different hospitals.

Risk Factors

The prevalence of risk factors is reported in Table 1. Analysis of the risk-factor distribution at individual hospitals showed similar percentages at all six.

Ortolani-Barlow Maneuver Results

The Ortolani-Barlow maneuver was done for the same baby by one or more pediatricians. In 134 cases (2.9%), the maneuver was carried out by one pediatrician; in 1762 cases (37.9%) by two; in 2706 cases (58.2%) by three; and in 46 cases (1.0%) by four. A positive test result was reported by one or more pediatricians in 233 subjects (5.1%). The Ortolani-Barlow maneuver gave a positive result in 128 (19.1%) of the 671 newborns in hospital four and in 105 (2.6%) of 3977 newborns in the other hospitals. When the positive results were expressed as a percentage of the total maneuvers carried out, values between 0.4 and 1.2% were obtained in the different hospitals—except in hospital four (4.9%). Thus, hospital four was excluded from the subsequent analyses of the Ortolani-Barlow maneuver.

In the subjects for whom the Ortolani-Barlow maneuver gave a negative result, clicks were identified in 2.2 to 13.4% of the maneuvers performed at the various hospitals.

Most of the positive maneuvers were not confirmed by the other pediatricians who carried out the test on the same baby. Of a total of 277 positive maneuvers, 227 (81.9%) were detected by one pediatrician alone, and in only 17 cases (6.1%) was the positivity confirmed by all the pediatricians.

TABLE 1. Prevalence of Risk Factors in Recruited Population

Risk Factors	No. of Cases	Prevalence (%)
Oligohydramnios	119	2.6
Breech presentation	210	4.5
Family history	469	10.1
Associated malformations	38	0.8

Sonography Screening Results

In 3509 patients (75.4%) the ultrasonography was performed within the first week of life, whereas in 1139 subjects (Lecco hospital) it was done at 22 days of life as a mean. The distribution of subjects by sonographic results is reported in Table 2. The subjects with type I bilateral hip constituted 50.1% of the population studied; type IIa, 44.8%; type IIc or IId, 4.5%; and type III or worse, 0.6%.

The distribution of the sonographic results varied considerably in the different hospitals; the frequency of a normal finding (type I) varied from 37.3 to 72.1%; that of type IIa, from 23.6 to 57.6%; that of type IIc-IId, from 0.8 to 7.0%; and that of type III-IV, from 0.0 to 1.1%.

Risk Factors and Sonographic Screening Results

The ratio between the presence of risk factors and the sonographic results is analyzed in Table 3. The relative risk (RR) is reported for subjects presenting with risk factors, calculated for three classes of sonographically demonstrated abnormalities: IIa, IIc to IId, and III to IV.

Comparison Between the Two Tests Used

Clinical agreement was evaluated considering the two test results as independent variables. The overall agreement of the tests in identifying worse than type IIa hip was 0.96, positive agreement 0.12, and the κ-statistic 0.11.

The accuracy of the Ortolani-Barlow maneuver was then calculated, with the sonographic result the gold standard. The sensitivity of the test in identifying subjects with type IIc-IId hip or worse was 0.07, the specificity 0.99, and the positive predictive value 0.35.

The relation between results of the Ortolani-Barlow maneuver and ultrasonography was also analyzed. The former result was considered a simple risk factor and the RR calculated (type IIa is deemed a degree of normality in this analysis). Subjects with a positive Ortolani-Barlow maneuver had a relative

TABLE 2. Distribution of Subjects by Sonographic Results

		Right Hip						Total
		I	IIa	IIc	IId	III	IV	
Left hip	I	2330	480	11	3	1		2825
	IIa	471	1130	43	12	5		1661
	IIc	16	55	25	3	3		102
	IId	6	9	13	12	4		44
	III		1	4	2	7		14
	IV			1			1	2
	Total	2823	1675	97	32	20	1	4648

TABLE 3. Relative Risk (RR) of the Different Risk Conditions

Risk	Degree	RR	CI	*P*
Female gender	IIa	1.29	1.21–1.37	.0000000
	IIc–IId	3.14	2.34–4.20	.0000000
	III–IV	8.18	2.85–23.42	.000002
Firstborn	IIa	1.03	0.97–1.10	.30
	IIc–IId	1.01	0.78–1.32	.98
	III–IV	1.36	0.65–2.87	.41
Oligohydramnios	IIa	1.03	0.84–1.24	.87
	IIc–IId	0.78	0.30–2.04	.78
	III–IV			
	IIa	1.10	0.96–1.27	.21
Presentation	IIc–IId	2.35	1.54–3.57	.0001
	III–IV	2.81	0.87–9.12	.1*
Family history	IIa	0.93	0.83–1.04	.21
	IIc–IId	2.14	1.56–2.92	.000003
	III–IV	3.35	1.50–7.48	.001

* Fisher's exact test.

risk of 7.2 (confidence interval [CI] 4.9–9.6) for type IIc-IId hip and 27.1 (CI 18.5–38.7) for type III-IV hip when compared to subjects with a negative maneuver. In subjects with clicks at clinical examination the relative risk was 2.1 (CI 1.4–2.5) for type IIc-IId hip and 1.8 (CI .5–6,2) for type III-IV hip.

Even when all the pediatricians agreed on a positive or negative clinical examination, discrepancies arose between clinical and sonographic results. In 15 cases of Ortolani-Barlow maneuvers were positive for all pediatricians, only in 6 did the sonographic examination reveal a pathologic picture (type III or IV); in 6 others the sonography showed an intermediate type (IIc or IId), and in 3 cases it did not detect a pathologic condition. In 7742 cases of agreement between all the pediatricians on a negative clinical examination, type III or IV hip was found by ultrasonography in 22 subjects and type IIc-IId in 206. A review of 22 false-negatives has demonstrated that the presence of asymmetric limbs or reduced abduction of the hip was not given due consideration during the examination. These conditions may occur in subjects with irreducible hip dislocations.

Follow-up of Subjects With Type IIa Hip

The screening examinations showed 2206 subjects (44.8% of the population) with a type IIa hip (bilateral or unilateral). A follow-up study was possible in 1927 (87.4%) subjects (range 73.3 to 95.8% for the individual hospitals). The mean age of the subjects at the first follow-up examination was 69 days.

Considering the number of hips (instead of subjects), 83.1% of type IIa hips at screening were normal at follow-up and 16% were confirmed as type IIa, whereas in 0.9% the sonographic result worsened. Follow-up of the subjects with monolateral type IIa hip demonstrated a worsening of the controlateral hip diagnosed at screening as type I in 3.4% of the cases.

DISCUSSION

Although the importance of DDH screening is widely recognized,[1–7,19] there is no definite agreement on the organizational model or the diagnostic screening tests that should be used. No population studies are yet available that meet the minimum requirements for correct execution and interpretation of the diagnostic (clinical and sonographic) tests and allow evaluation of clinical screening results done in different hospitals. Our study, performed at six hospitals in which clinical and sonographic DDH screening procedures were already under way, represents a concrete example of screening results carried out at hospitals typical of the Italian national health system.

We also studied some previously suggested risk factors[6,12,20] and found an association between type III or IV hip and female gender (RR = 2.8), family history of DDH (RR = 3.4), and breech presentation (RR = 2.8). A weaker association with type IIc and IId hip was also detected.

Analysis of the Ortolani-Barlow maneuver results was complex because, according to the design study, the maneuvers were carried out on the same subject by several pediatricians. Excluding the hospital in which the frequency of positive maneuver was particularly high, the ratio of subjects with positive Ortolani-Barlow tests was 15/1000, a figure similar to that reported by other authors.[7,21,22]

The reproducibility of the Ortolani-Barlow maneuver was the main problem to emerge from our study. Wide variations in the frequency of detection of positive maneuvers and clicks were also observed among the different hospitals; this could be explained as a different prevalence of the disorder, but in view of the homogeneous recruited population, the variations were probably attributable to incorrect execution of the clinical examination.

The inconsistent results obtained by the pediatricians who made the clinical investigations show clearly that the Ortolani-Barlow maneuver, as carried out by the six hospitals, cannot be considered a reliable screening test. We believed that clinical training of two doctors from each hospital in a specialized center and clinical practice with a simulator for the other doctors would make the test sufficiently reliable, but this was not the case. In our opinion the low reproducibility of the clinical investigation was due not to unreliability of the maneuver itself but to insufficient clinical training of the investigators. This was confirmed by the admission of most of the physicians who attended the specialized center that they were not sufficiently expert to carry out an accurate clinical examination of the hip. Other studies have shown that highly skilled personnel are needed to obtain good results from clinical screening, and this is possible when only two or three physicians of the staff perform the clinical test.[23] Good results can be obtained also by nonmedical investigators, provided they are thoroughly trained to carry out the clinical test.[24]

The reliability and interobserver variability between ultrasound examination at birth (very unreliable) and at 4 weeks (more reliable) vary considerably. In our study ultrasonography was performed in 75.4% of cases within the first week of life.

According to the results of the neonatal ultrasound screening examination, only 50.1% of the studied population could be described as normal; of the subjects examined, 44.8% had a type IIa hip (unilateral or bilateral), which inevitably requires a repeat sonography and, according to several authors, postural treatment.[16,25] Type IIc or IId hip(s) were found in 43/1000 subjects, and type III or IV in 6/1000 of our population. The latter figure does not differ significantly from those reported in the literature.[16–22] Considerable variations were also seen in the data from different hospitals relative to both sonographic and clinical examination.

Analysis of the relationship between the results of the Ortolani-Barlow maneuver and the sonographic examination is difficult due to the lack of a valid gold standard. If sonography is assumed as a gold standard, as is done to some extent in our study, this brings with it the risk of an excessive number of diagnoses. Analysis of DDH prevalence reported in the literature shows a constant increase each time a new test is used.[2,12,26] However, the fact that, for ethical reasons, all patients with a positive result undergo treatment independently of the test used impedes evaluation of the specificity of the test. The detection of false-negatives was difficult, and it cannot be excluded that arthrosis of the hip in adulthood is not due to an undetected abnormality present in neonatal age.[27]

Bearing in mind these methodologic limits, the results of our study demonstrate that if sonography is assumed to be the gold standard and if sonographic results worse than type IIa are considered pathologic, the sensitivity and positive predictive value of the Ortolani-Barlow test are extremely low. Positive Ortolani-Barlow maneuver was associated significantly with a sonography showing a type worse than IIa; the RR was statistically significant ($P < .00001$).

The ultrasound detection of a type IIa hip raises screening costs considerably, in that half of the population must undergo a further sonographic examination, which, as shown by our follow-up data, very often shows a normalization of the clinical picture. Screening done at 60 days of life would reduce the number of subjects with type IIa hip(s) (requiring repeat sonography) by 78 and 64%, respectively, in the case of unilateral and bilateral involvement. This would result in a considerable reduction in the screening costs.

An improvement in screening efficiency could be achieved by screening subjects with risk factors (family history, breech presentation, positive Ortolani-Barlow maneuver) in the neonatal period.[12] All other subjects could be screened at 60 days of life. About 50% of the subjects with dislocation (type III or IV hip) and 34% of those with type IIc-IId hip could be detected by screening 15% of the population in the neonatal period, and therapy could be anticipated. Obviously a strategy that delays screening to a later age than the neonatal period could in theory increase the number of babies lost to screening. Based on our experience, such losses can be minimized by a careful information program addressed to the families.

To lower DDH screening costs, some authors[10–12] have proposed that only subjects with risk factors or positive clinical examination should undergo sonographic screening. To evaluate this proposal, we applied these restrictions on the examined population. Had we screened all females and the males with a risk factor (family history, breech or transverse presentation, positive clinical examination), there would have been a population of 2730 subjects (instead of 4648) and a loss of 7% in diagnosis of subjects with sonography showing type III or IV hip and of 15% for type IIc or IId hip.

In conclusion, our results demonstrate that some aspects of DDH screening need further detailed study; the reduced reproducibility of the diagnostic tests used in screening is the main problem. The study was conducted in a relatively small geographic area that was ethnically homogeneous. This makes it improbable that the observed differences are due to a different prevalence of DDH. The insufficient agreement between pediatricians who carried out the Ortolani-Barlow maneuver indicates that this examination, as now performed, cannot be proposed as a screening test. Specific training given to the doctors participating in the study was not sufficient to warrant acceptable reproducibility of the test. It was not possible to evaluate the reproducibility of the ultra-

sound examination in this study. The substantial differences observed among the different hospitals lead us to hypothesize that important problems of reproducibility exist even in these cases. A correct DDH screening model for Italian hospitals cannot therefore be defined at this time.

APPENDIX

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REFERENCES

1. Ortolani M. *La lussazione congenita dell'anca*. Bologna, Italy: Cappelli Editore; 1948
2. Editorials. Screening for congenital hip dysplasia. *Lancet*. 1991;1:947–948
3. Hansson G, Nachemson A, Palmén K. Screening of children with congenital dislocation of the hip joint on the maternity wards in Sweden. *J Pediatr Orthop*. 1983;3:271–279
4. Screening for the detection of congenital dislocation of the hip. *Arch Dis Child*. 1986;6:921–926
5. Tredwell SJ, Bell HM. Efficacy of neonatal hip examination. *J Pediatr Orthop*. 1981;1:61–65
6. Hensinger RN. Congenital dislocation of the hip: treatment in infancy to walking age. *Orthop Clin North Am*. 1987;18:597–602
7. Dunn PM, Evans RE, Thearle MJ, Griffiths HE, Witherow PJ. Congenital dislocation of the hip: early and late diagnosis and management compared. *Arch Dis Child*. 1985;60:407–414
8. Graf R. The diagnosis of congenital hip joint dislocation by the ultrasound compound treatment. *Arch Orthop Trauma Surg*. 1980;97:117–33
9. Graf R. New possibilities for the diagnosis of congenital hip joint dislocation by ultrasonography. *J Pediatr Orthop*. 1983;3:354–59
10. Clarke NMP, Clegg J, Al-chalabi A. Ultrasound screening of hips at high risk for CDH: failure to reduce the incidence of late cases. *J Bone Joint Surg*. 1989;71:9–12
11. Rosendahl K, Markestad T, Lie RT. Congenital dislocation of the hip: a prospective study comparing ultrasound and clinical examination. *Acta Paediatr*. 1992;81:177–181
12. Walter RS, Donaldson JS, Davis CL, Shkolnik A, Binns HJ, Carroll NC, Brouillette RT. Ultrasound screening of high risk infants. A method to increase early detection of congenital dysplasia of the hip. *Am J Dis Child*. 1992;146:230–234
13. Exner GU. Ultrasound screening for hip dysplasia in neonates. *J Pediatr Orthop*. 1988;8:656–660
14. Berman L, Klenerman L. Ultrasound screening for hip abnormalities: preliminary findings in 1001 neonates. *Br Med J*. 1986;293:719–722
15. Szoke N, Kuhl L, Heinrichs J. Ultrasound examination in the diagnosis of congenital hip dysplasia of newborns. *J Pediatr Orthop*. 1988;8:12–16
16. Tonnis D, Storch K, Ulbrich H. Results of newborn screening for CDH with and without sonography and correlation of risk factors. *J Pediatr Orthop*. 1990;10:145–152
17. Ortolani M. Un segno poco noto e sua importanza per la diagnosi precoce di prelussazione congenite dell'anca. *Pediatria*. 1937;2:129–133
18. Barlow TJ. Early diagnosis and treatment of congenital dislocation of the hip. *J Bone Joint Surg*. 1982;44b:292–302
19. Aronsson DD, Goldberg MJ, Kling TF Jr, Roy DR. Developmental dysplasia of the hip. *Pediatrics*. 1994;94:201–208
20. Dunn PM. Perinatal observations of the etiology of congenital dislocation of the hip. *Clin Orthop*. 1976;119:11–22
21. Burger BJ, Burger JD, Bos CFA, Obermann WR, Rozing PM, Vandenbroucke JP. Neonatal screening and staggered early treatment for congenital dislocation or dysplasia of the hip. *Lancet*. 1990;2:1549–553
22. Atti G, D'Eramo C, Baronciani D. Diagnosi di lussazione congenita dell'anca: confronto tra esame clinico ed indagine ecografica. *Riv Ital Pediatr*. 1192;18:612–618
23. Sommerville EW. The clinical diagnosis of congenital dislocation of the hip. *Curr Orthop*. 1987;1:255–257
24. Bernard AA, O'Hara JN, Bazin S, Humby B, Jarret R, Dwyer JP. An improved screening system for early detection of congenital dislocation of the hip. *J Pediatr Orthop*. 1987;7:277–282
25. Graf R. *Guide to Sonography of the Infant Hip*. Stuttgart, Germany: Georg Thieme Verlag; 1987
26. Rosendhal K, Markestad T, Terje Lie R. Ultrasound screening for developmental dysplasia of the hip in the neonate: the effect on treatment rate and prevalence of late cases. *Pediatrics* 1994;94:47–52
27. Bennet JT, MacEwen GD. Congenital dislocation of the hip. Recent advances and current problems. *Clin Orthop*. 1989;247:15–21

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DOI: 10.1542/peds.99.2.e5

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