

REVIEW

Paediatric imaging

Ultrasonography in developmental dysplasia of the hip: A review of current clinical strategies and recommendations for revision of practice

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ABSTRACT

Developmental dysplasia of the hip is a broad term covering a wide spectrum of hip joint disorders, ranging from maturation deficits to severe dysplasia or dislocation. Published prevalence of the disorder ranges from 0.25% to 2.5% or even more in certain geographic areas. Risk factors do exist and include female gender, white race, positive family history and mechanical restriction during or after birth. Low sensitivity and specificity of clinical examination promoted the

development of several sonographic techniques for early diagnosis. Among the above-mentioned techniques, Graf's technique, supported by extended literature and epidemiological data, offers an anatomically based description of pathology and effective monitoring of treatment. Universal sonographic screening early in life is strongly recommended and initiation of treatment as early as possible is mandatory for an optimal outcome.



KEY WORDS

congenital; dislocation; hip joint/dysplasia; ultrasonography/diagnosis; infant



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Introduction

Developmental dysplasia of the hip (DDH) is a condition that includes a wide spectrum of disorders, which share the potential of causing significant long-term complications that may lead to severe disability during early adult life.

Most of the cases of DDH are potentially reversible if diagnosed and treated early. Therefore, the role of imaging, which is the mainstay of early diagnosis, is critical. Especially ultrasonography (US), the imaging modality of choice, has evolved to be the one-stop-shop imaging method for early diagnosis of DDH and the preferred tool for treatment evaluation and monitoring.

In this manuscript, we sought to review the published literature, focus on the pros and cons of US, present current clinical practice and propose an algorithm for screening.

Definition of DDH

DDH is the term which has replaced congenital dislocation of the hip in medical literature. This is mainly because it covers a wider spectrum of disorders, ranging from maturation deficits to severe dysplasia or dislocation [1, 2]. Moreover, it better describes a condition with a poorly understood natural history, the origin of which is both congenital and developmental.

The existence of various approaches to infant hip assessment, some of them being clinical and others being imaging (mainly sonographic with various techniques), makes the effort of defining pathology even more complicated, leading to several variations [2]. Fortunately, with the use of US, the exact anatomy of the infant hip joint has nowadays been studied thoroughly, facilitating a systematic approach to the joint disorders.

The definition of DDH does not include hip joint disorders which are due to co-existing medical conditions (cerebral palsy, slipped capital femoral epiphysis, Legg-Calve-Perthes disease etc.), teratologic disorders and cases of traumatic hip dislocations [3, 4].

Epidemiology

Depending on whether we refer to the pre- or the post-sonographic screening era, prevalence of DDH in the literature ranges from 0.07% to 0.15% and 0.25% to 2.5% (or even higher in certain geographic areas) respectively [3, 5, 6]. Significant geographic variability is due to true population differences, but also due to the defini-

tion variations mentioned above [7-9]. In most DDH cases, up to 80% in published reports, the disorder affects the left hip joint. This has been attributed to fetal positioning (left occiput anterior), with the left side of the fetus being more restricted and adducted as it is adjacent to the maternal sacrum, especially during the 3d trimester of pregnancy [10]. More important than the exact prevalence of the disorder is the fact that almost 1/3 of total hip replacements in patients younger than 60 are due to undiagnosed or untreated cases of DDH [11], most of which (95-98%) might have the chance of being treated if diagnosed earlier.

Risk factors

Risk factors according to the literature [12-15] include female gender, white race and positive family history of DDH. Mechanical restriction during pregnancy (oligohydramnios, breech presentation, macrosomic babies), during delivery (vaginal birth in breech presentation with extended legs) or after birth (swaddling), significantly contribute to morbidity. On the contrary, lack of mechanical charge explains lower incidence of DDH in premature babies. Presence of risk factors alerts the clinician to perform a thorough clinical/imaging examination, although most of the DDH cases are diagnosed in babies without any identifiable risk factors. So, it is clear that DDH screening based solely on risk factor identification is not justified [2, 16].

Clinical vs. Imaging Examination

Physical examination of the hips is the initial step of hip assessment. It is usually carried out at birth by the neonatologist and subsequently routinely by the paediatrician [17]. Clinical examination mainly consists of provocative tests (namely Ortolani and Barlow (**Fig. 1**), assessment of joint motion range, measurement of leg length and assessment of skin fold symmetry. Although it remains the mainstay of screening in several health systems and guidelines, it is neither specific nor sensitive and requires careful consideration and clinical experience [18, 19]. Dysplastic hips may remain clinically silent when the femoral head is centered [20, 21]. Even cases of severe hip dysplasia may remain clinically obscure [12, 21, 22]. The combination of risk factors and clinical history may perform slightly better; however, it is still insufficient to provide an effective screening method.

US offers us the opportunity to image the non-os-

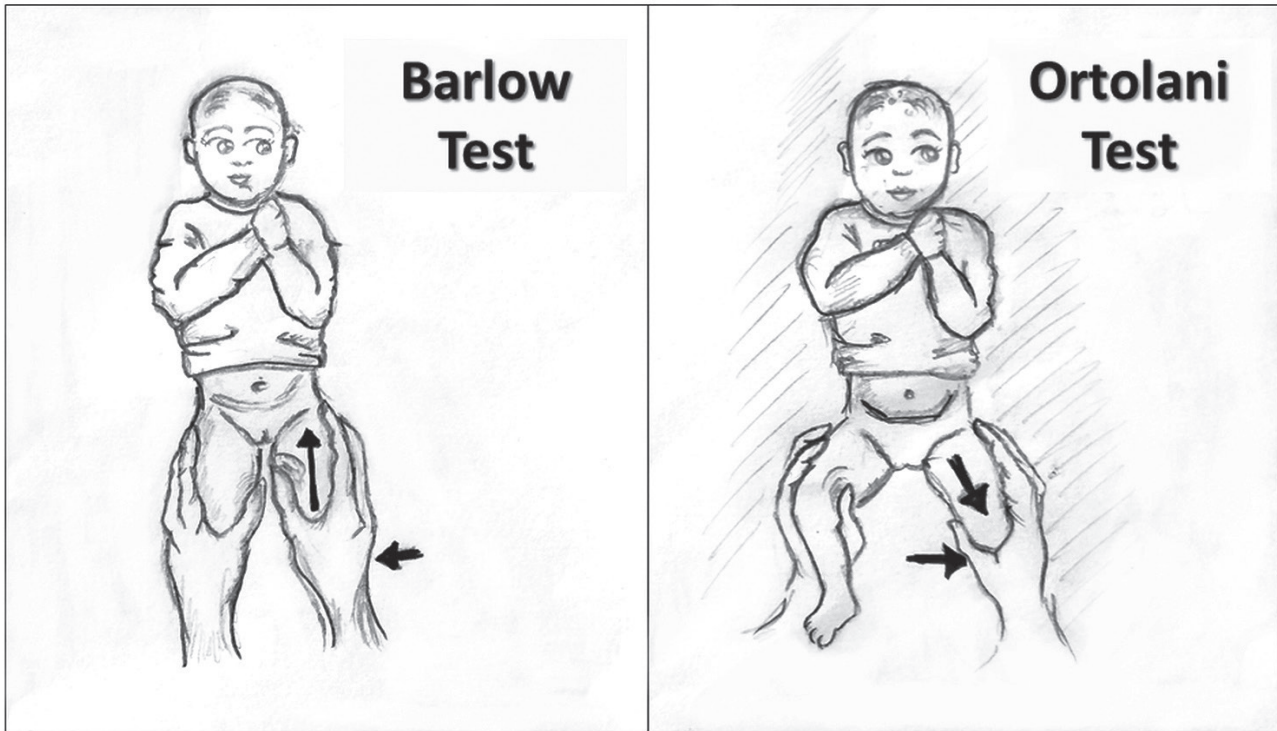


Fig. 1. Barlow and Ortolani maneuvers consist part of the standard infant examination performed by the pediatrician in early infancy. Barlow maneuver aims to identify unstable hips and consists of the application of a posterior force with the hip adducted, which produces a posterior dislocation of the joint (with a palpable clunk). On the contrary, Ortolani maneuver consists of the application of anterior traction on an abducted / flexed dislocated hip, which reduces (with a palpable clunk) the dislocation

sific(-ied) parts of the hip joint very early in life. Cartilaginous structures (femoral head, cartilaginous roof, labrum), joint capsule and the muscles are adequately examined with US, both in an anatomic and dynamic (when needed) way (Fig. 2). At the same time, integrity and adequacy of the acetabular bony roof is assessed, both qualitatively and quantitatively, and the position of the femoral head within the joint socket is documented. Several different sonographic techniques have been developed and clinically assessed, a few of which are still in clinical practice [23-27]. X-ray nowadays has a historical role in DDH screening and is preserved mainly for late presenting cases and treatment monitoring when US is no more technically feasible [28]. CT and MRI still have a role when examining the consequences of neglected or maltreated cases of DDH or when planning hip surgery [29].

Hip US

The era of hip US began in 1980 when Reinhard Graf published the original paper about hip US for the diagnosis of congenital dislocation of the hip in infants [23].

Over the next years, several publications suggested modifications or advances of this technique, or different techniques altogether. Traditionally, examination techniques have been categorised according to their imaging focus (acetabular morphology, femoral head coverage) or their technical approach (static vs. dynamic). It is beyond the scope of this paper to describe every different sonographic technique that is or has been used or proposed. It is our aim to emphasise the main advantages and disadvantages of the most widely used ones and then conclude with evidence based conclusions / recommendations.

Femoral head coverage

Measuring the femoral head coverage (FHC), from a technical point of view, seems to be the easiest and more reproducible way to assess a hip joint. On a standard coronal hip scan the percentage of the femoral head covered by the acetabular roof is calculated as demonstrated on the figure (Fig. 3). There are however certain methodological problems that significantly reduce the value of

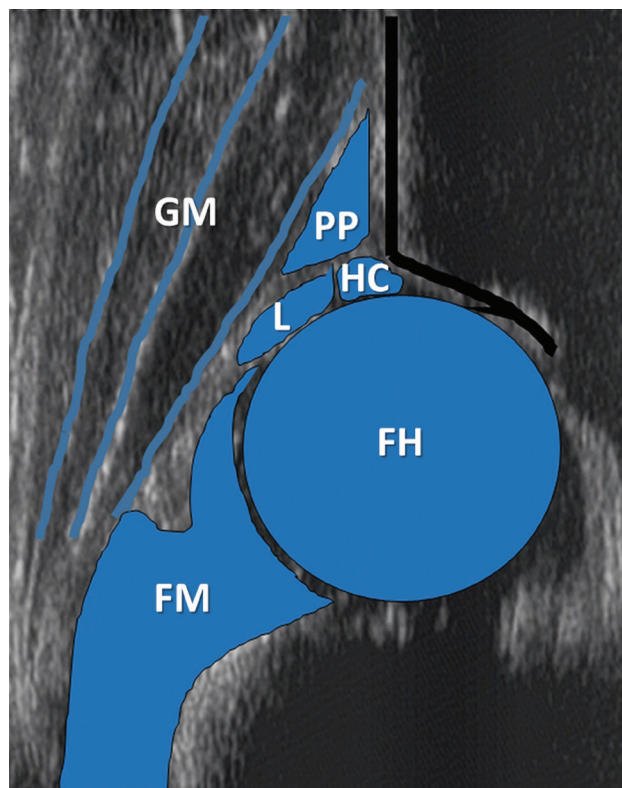


Fig. 2. Cartilaginous structures (femoral head, cartilaginous roof, labrum), joint capsule and the muscles are adequately examined with ultrasound (blue colour). FH: Femoral Head, FM: Femoral Metaphysis, L: Labrum, HC: Hyaline Cartilage, PP: Proximal Perichondrium, GM: Gluteal Muscles / Intermuscular Septa

the technique, which was originally proposed by Morin (USA) and Terjesen (Norway) [25, 26] and is still utilised in Northern Europe. Ovoid shape (and not spherical) of the femoral head [30], alongside with failure to define a standard plane for measurement, makes the method vulnerable to rotational distortions. Furthermore, the classification scheme utilised for assessment has a wide “gray” zone (FHC<50% may be abnormal) and an unsound reasoning behind it. As a result, it is of limited use, restricted in certain geographic areas [31].

Hip Stability - Dynamic assessment

Dynamic assessment of the hip joint was introduced by Harcke and coworkers in USA and mirrored the hypothesis that correct development of the acetabulum was heavily dependent on the position of the femoral head [24, 32]. Examination includes static and dynamic evaluation of hip stability in two examination planes (coronal and transverse) with the baby lying supine or lateral,

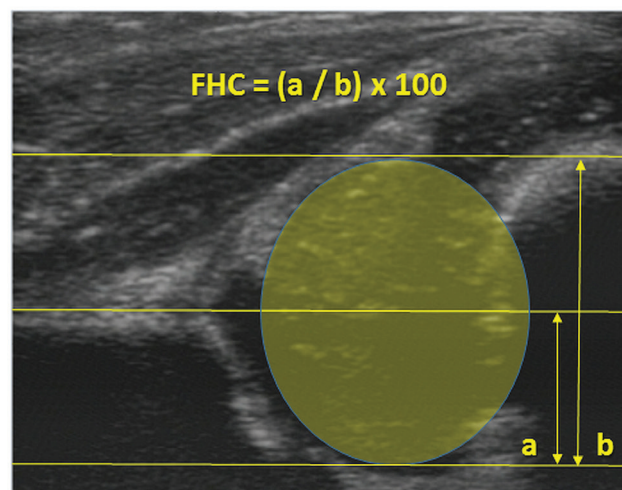


Fig. 3. Assessment of Femoral Head Coverage (FHC). FHC (percentage) is calculated in a standard coronal scan by dividing acetabular width (distance a, measured from the medial part of the acetabulum to a line parallel to the iliac bone) by the femoral head diameter (distance b, measured between lines parallel to the iliac line, the first from the medial part of the acetabulum as in distance a and the second from the outer part of the cartilaginous femoral head). FHC is calculated by the formula $a/b \times 100\%$

and the hip in both neutral and flexed position (dynamic four-step method). A modified Harcke technique (including an optional rough assessment of the acetabular morphology and femoral head coverage) is the technique currently proposed by the American Institute of Ultrasound in Medicine, developed in conjunction with the American College of Radiology, the Society for Paediatric Radiology and the Society of Radiologists in Ultrasound [33].

The main drawbacks of the aforementioned “dynamic” examination techniques are the following: Examination methodology (free hand, not fixed baby position) may cause certain technical problems with image acquisition, consisting mainly of rotational / tilting effects that may remain undetected (due to limited scanning plane definition) and may heavily affect image diagnostic quality and interpretation. Examination in a transverse plane does not offer any additional information and does not solve any diagnostic problems. Furthermore, qualitative

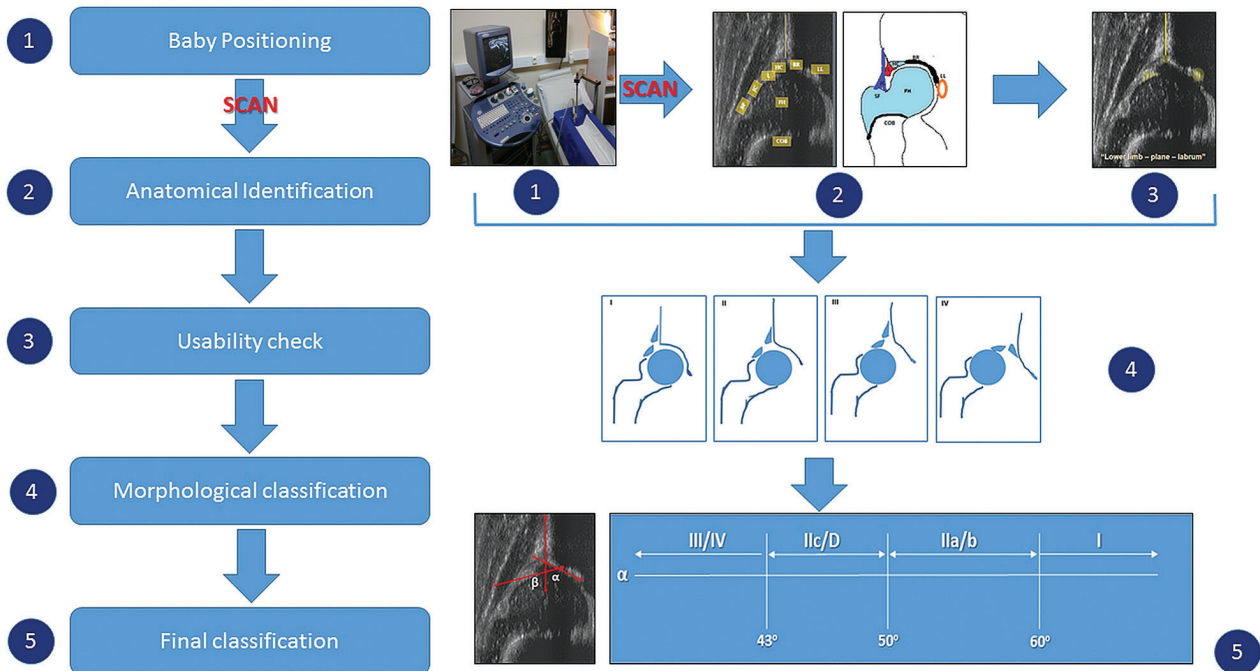


Fig. 4. Graf's technique offers a well-structured anatomical approach, based on the application of a stepwise, strictly defined protocol

interpretation of the examination results and the proposed simplified hip joint classification may be a cause of concern when following up hip treatment and a cause of subjectiveness of the results, especially when comparing the results of different examiners. Arbitrary (or on demand) addition of extra examination bits (including acetabular assessment and femoral head coverage) is not acceptable for a universal screening test which must follow a strict examination protocol. Finally, the dynamic part of the sonographic examination may be very uncomfortable and irritating for the baby (if not hazardous under certain circumstances), is not justified in the majority of cases where acetabular morphology is normal (a high correlation has been shown between acetabular morphology and hip stability in several publications) and hip instability is not separated objectively from harmless movements.

Graf's technique

The technique that is utilised in most European countries is Graf's technique. The main advantage of this technique is that it offers a well-structured anatomical approach, based on the application of a stepwise, strictly

defined protocol (Fig. 4). The addition of specified checklists for the examiner helps in avoiding certain technical mistakes and further enhances correct interpretation and classification in certain categories. The examination is carried out in the lateral position using an examination cradle with a fixed probe guide. This setting is considered a necessary adjunct for correct image acquisition and maintenance of a standard examination plane. Image acquisition is followed by anatomical identification. The acquired image is only acceptable for diagnosis if it depicts certain anatomic structures. If any of them is missing, then it is considered unacceptable and a new image must be acquired (Fig. 5). Usability check follows anatomic identification. As already mentioned, it is absolutely necessary to define a standard plane of examination, because only then can the examiner be confident that the image is correctly acquired, lines and angles can be drawn and the examination is appropriate for clinical judgment. For this purpose, a standardised approach is utilised (lower limb-plane-labrum) to define the correct scanning position and plane (Fig. 6).

Provided that a correct image at a standard scanning plane has been acquired, morphological classification

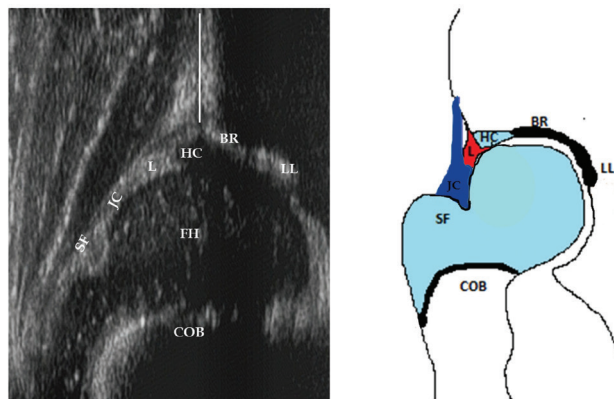


Fig. 5. Right Hip, Coronal Plane – Anatomic identification: COB – ChondroOsseous Border, FH – Femoral Head, SF – Synovial Fold, JC – Joint Capsule, L – Labrum, HC – Hyaline Cartilage, BR – Bony Roof, LL – Lower Limb os ilium

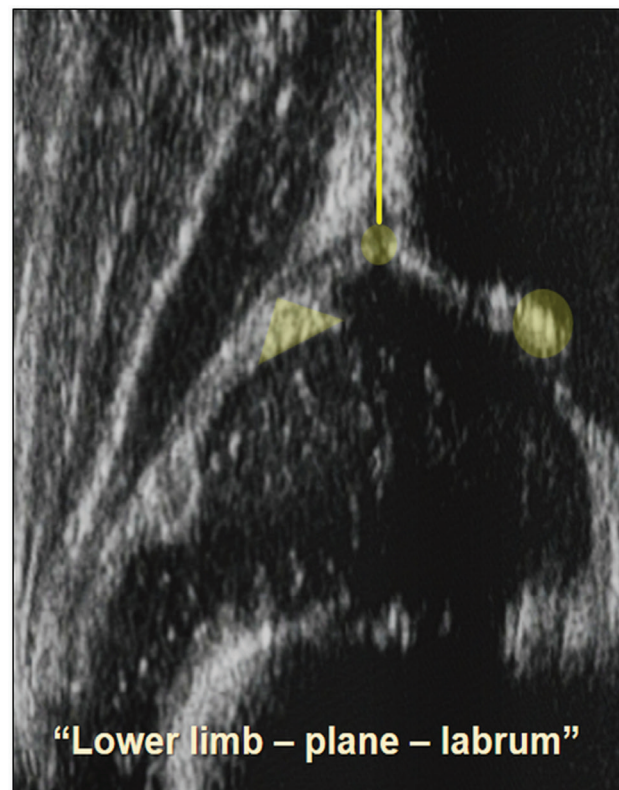


Fig. 6 (right). Defining the Standard Plane of examination. For this purpose, a standardised approach is utilised (lower limb–plane–labrum) to define the correct scanning position and plane. The yellow triangle represents the labrum, the straight line the correct scanning plane and the yellow circle the lower limb of the os ilium

into four main categories (I to IV) is then performed and comprises the first step of the interpretation of the scan (Fig. 7a, b). It is mandatory that subjective morphological assessment is in full accordance with the objective classification with the use of measurements. Acetabular morphology is described with the use of two angles, the α angle which is the angle of acetabular inclination (bony roof angle) and the β angle which is the cartilaginous roof angle. Each hip is classified into one of four main types (nine subtypes) and clinical management is based on this classification (Table 1).

Hip examination and type classification is performed in a resting position (without stress). A dynamic examination is only preserved for a specific hip joint category (IIc). In those cases, separating harmless movements from (pathological) instability is important for treatment decisions. The same examination protocol and hip type categorisation is also used for treatment monitoring. Strict adherence to the above-mentioned examination steps (correct baby positioning, anatomic identification, usability check etc.) eliminates the risk of mistakes. However, there are cases where poor examination technique, no usability check, wrong anatomic identification or incorrect measurements have led to misdiagnoses.

Graf's examination technique's main criticism is about its complexity. This is mainly due to the fact that it no longer uses the original clinical and x-ray classification of normal, dysplastic, subluxed and dislocated hip, but classifies hip joints according to the exact anatomic pathology which must be identified and treated appropriately. What is considered by many the main disadvantage of the technique is actually its main strength: on the basis of this classification, treatment can be conducted appropriately according to the exact sonographic hip type. Monitoring of treatment is much easier and more objective since evaluation is no longer qualitative [34].

Selective vs. Universal Screening

Although universal clinical screening has been common clinical practice for many decades, universal hip screening has not gained general approval [2, 35]. The reason for this is mainly the fact that there is wide geographic variation in screening policies, published literature is heterogeneous (screening methods and population composition differ in different studies) and there are no randomised trials (it is not ethical to perform any). The fact that there are different diagnostic definitions for DDH further complicates the situation. Thus, at least at first

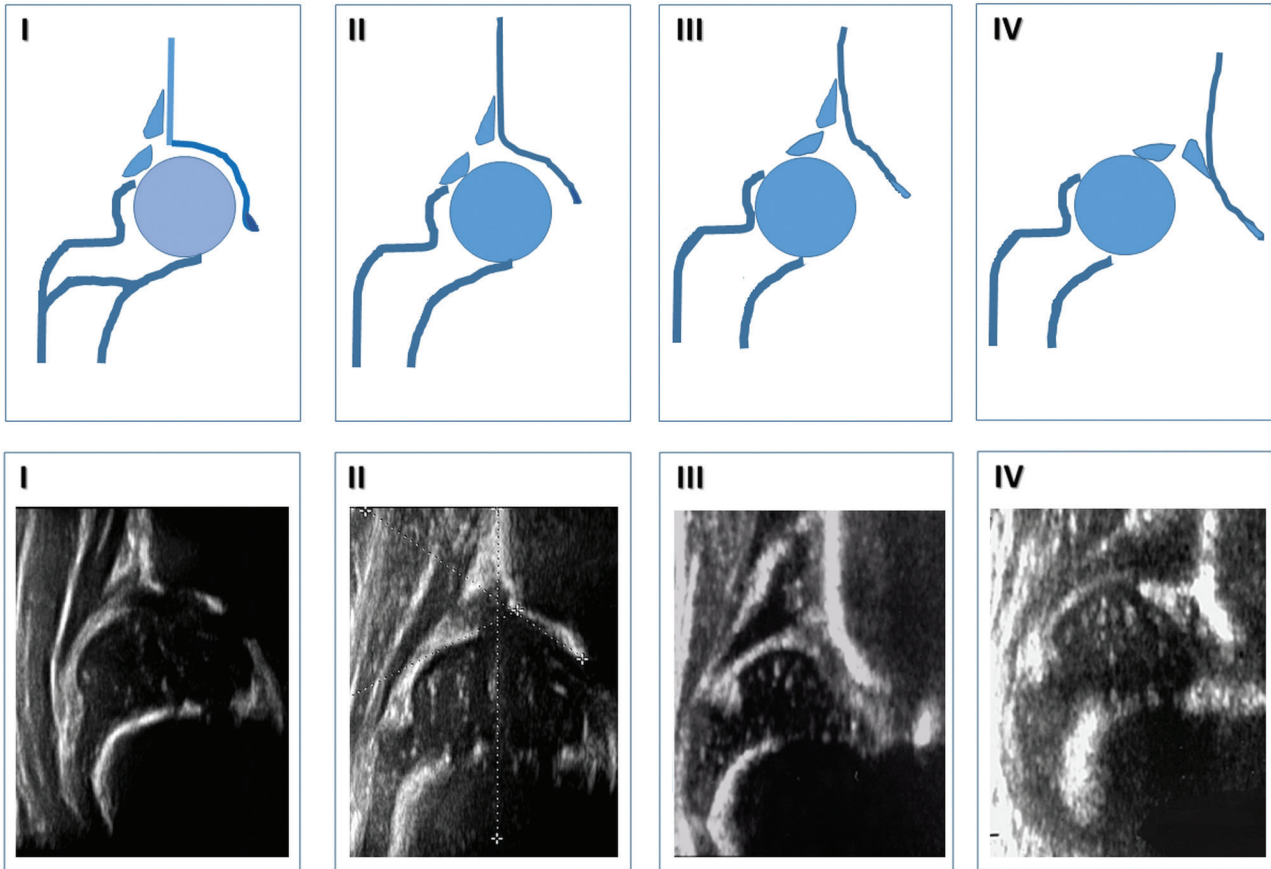


Fig. 7a (above), 7b (below). Provided that a correct image at a standard scanning plane has been acquired, morphological classification into four main categories (I to IV) is then performed. This comprises the first step of the interpretation of the scan

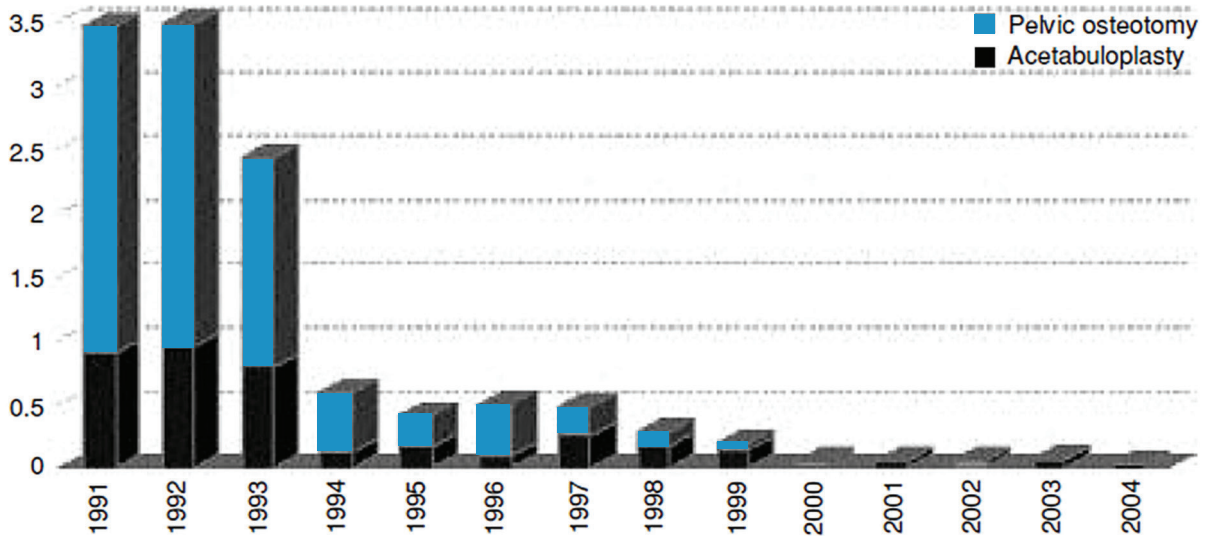


Fig. 8. Dramatic drop of surgical intervention rate in young children documented in Austria between 1991 and 2004 (used with permission: Graf R. The use of ultrasonography in developmental dysplasia of the hip. Acta Orthop Traumatol Turc 2007; 41 Suppl 1: 6-13)

Table 1. Hip type classification according to Graf

Hip Type (Graf)	Bony roof / α angle	Cartilagenous roof / β angle	Bony rim	Age
I / Mature Hip	Good / $\alpha \geq 60^\circ$	Covers femoral head Ia ($\alpha < 55^\circ$) / Ib ($\alpha > 55^\circ$)	Angular / blunt	Any age
IIa / Physiologically immature	Adequate $\alpha = 50-59^\circ$	Covers femoral head	Rounded	0 – 12 weeks
IIb / Delayed ossification	Deficient $\alpha = 50-59^\circ$	Covers femoral head	Rounded	> 12 weeks
IIc / D	Severely deficient $\alpha = 43-49^\circ$ or decentering (D)	Covers femoral head ($\beta < 77^\circ$) / displaced ($\beta > 77^\circ$)	Rounded to flattened	Any age
III Decentered hip	Poor / $\alpha < 43^\circ$	Pressed upwards	Flattened	Any age
IV Decentered hip	Poor / $\alpha < 43^\circ$	Pressed downwards	Flattened	Any age

sight, it seems impossible to collect high quality evidence to support universal sonographic screening.

On the other hand, in Austria and Germany, universal population screening with US is being carried out since the 1990's and there is sufficient epidemiological data to support it [35-37]. Dramatic decrease of treatment costs and surgical intervention rates in young patients with DDH were shown (Fig. 8, 9). The facts that most DDH cases affect low risk populations and that screening performance based on clinical examination and risk factors, as mentioned above, is suboptimal, further enhance the opinion that all infants should be sonographically screened for DDH.

A significant number of publications which are critical of Graf's technique, supporting clinical over sonographic screening for DDH, exhibit images which are suboptimal and thus non-diagnostic (according to the stepwise examination protocol presented above). This further supports the attitude that high quality hip US is not always available and proves that training has not been as optimal as desired. Erroneous US diagnoses are common and are mainly due to suboptimal technique. According to Graf, bedside teaching by non-authorized teachers must be rejected because it promotes habitual faults being passed on [38, 39]. Continuous advancement of hip US necessitates extra tuition of instructors. Thus, it is mandatory, in parallel with

the screening policy, to adequately train the examiners and maintain a high level of competence by a carefully designed certification and audit scheme.

Timing of Screening

Appropriate US screening time has been a matter of debate. Maturation of the hip joint (measured as an increase of the α angle), is more rapid during the first few months of life [40]. Therefore, scanning babies as early as possible is important in order to achieve the best results of treatment, if needed. To eliminate unnecessary re-examinations, it would be reasonable to perform US screening early enough to provide the best possible outcome in treated cases, while at the same time offering an appropriate time window for the spontaneous resolution of immature hips. Performing a scan during the first week of life in clinically unstable hips or high-risk cases and screening all babies between the fourth and sixth week might be an optimal screening protocol [41]. However, any policy that would involve universal US screening until the second month at the latest would be sufficient.

Selection of the appropriate screening scenario is heavily dependent on local or national circumstances [37, 38] and the aim would be to ascertain that all babies would be scanned at some time during the first weeks of life.

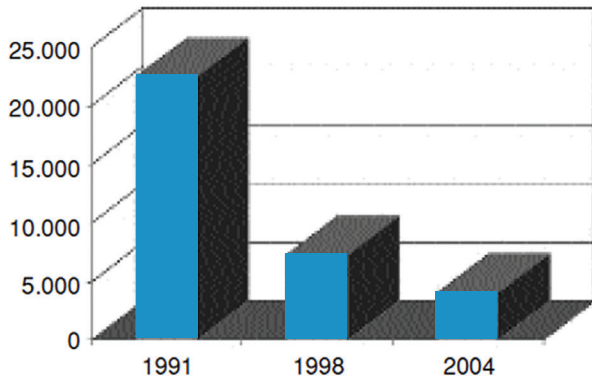


Fig. 9. Dramatic drop of treatment costs of DDH in young children documented in Austria between 1991 and 2004 (used with permission: Graf R. The use of ultrasonography in developmental dysplasia of the hip. *Acta Orthop Traumatol Turc* 2007; 41 Suppl 1: 6-13)

Management of DDH according to Graf [40]

DDH treatment aims at (1) centering the head within the acetabulum to facilitate correct development of the former, thus preventing future limping and (2) eliminating acetabular dysplasia, thus preventing early onset of arthritis and the need of early arthroplasty in a young adult. Treatment algorithm consists of femoral head reduction, maintenance of head relocation (retention) and correction of any residual acetabular dysplasia (maturation).

There is still some controversy about the correct timing of initiation of treatment (especially head reduction). There is some concern that reduction maneuvers should be performed after the ossification nucleus appears, in order to avoid head necrosis. According to the majority of published reports, early initiation of treatment is suggested.

The key in reduction technique is flexing the hips at 110° with a maximum abduction of 45°. Correct positioning is of utmost importance, to avoid causing avascular

necrosis of the femoral head due to exertion of excessive pressure on the centered head.

Treatment of dislocated or unstable dysplastic hips consists of:

- femoral head reduction: in babies younger than 6 months, early (as soon as possible) closed reduction of the femoral head / in older babies, open reduction (except when the femoral head is relocating easily),
- application of a spica cast for 4-6 weeks (retention), although in newborns some centers may use the Pavlik harness,
- depending on the age of the baby, a Pavlik harness is used until the hip joint turns to a Graf Type I hip (maturation) or until the baby is too old for a Pavlik or other flexion - abduction device (or plaster). If there is a residual dysplasia, a pelvic osteotomy is arranged at the age of 2 years or later.

In stable (centered) dysplastic hips, therapy begins from the maturation step (c).

Conclusion

US offers an effective and accurate method for the early diagnosis of DDH. Among the most popular sonographic techniques, Graf's technique is a powerful screening tool which is based on anatomical description of hip pathology, and combines early diagnosis with effective monitoring of treatment. Its universal application for more than two decades in central European countries has been clinically useful and thus it is generally recommended. Timing of screening should be modified according to regional peculiarities. It is however mandatory that diagnosis should not be made later than eight weeks to ensure early initiation of treatment. The treatment algorithm consists of femoral head reduction, maintenance of head relocation (retention) and correction of any residual acetabular dysplasia. **R**

Conflict of interest:

The authors declared no conflicts of interest.

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