REVIEW

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Hip Sonography for Developmental Dysplasia of the Hip: The Graf Method

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ABSTRACT

Developmental Dysplasia of the Hip (DDH) is one of the most common musculoskeletal disorders for children. Multiple risk factors as female sex, positive family history, breech positioning in utero are blamed for the presentation of this entity. The hip joint of the infant is affected and more specifically, the acetabulum remains shallow and the femoral head grows in a wrong position. Until 1980 the diagnosis was based on clinical examination (Barlow and Ortolani maneuvers) and radiological imaging. The diagnosis was not certain and many cases remained undiagnosed until childhood or adolescence. In 1980 Prof. Reinhard Graf presented his ultrasonographic method for the examination of the infant hip joint. This method evaluates the bony and cartilaginous coverage of the femoral head by the acetabulum in the infantile joint by measuring α-angle and β-angle. Using and valuating these measurements the hip joint is later on classified by Graf classification (Types I-IV) and each type corresponds to a specific treatment option. Graf method is the most used ultrasonographic examination in many European countries and has contributed to the reduction of neglected DDH cases and decreased the number of complex pelvic operations that children with dysplastic hips should undergo to be cured.

KEYWORDS: Developmental Dysplasia of the Hip (DDH), Graf method, hip ultrasonography, preventive screening

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Introduction

Developmental Dysplasia of the Hip (DDH) is a common musculoskeletal disorder for children, with a median prevalence of 0.1–2 per 1,000 infants.[1] According to the World Health Organization (WHO) 10%-15% of patients with hip replacement under the age of 50 have positive clinical history of infant hip dysplasia.[2]Developmental dysplasia of the hip (DDH) describes the malformation of hip joints in newborns. This disorder contains a wide spectrum of conditions from acetabular dysplasia, to severe dysplasia with dislocation and possible irreducible hip dislocation with proximal femoral displacement.[3] DDH defines congenital malformation, other perturbations during development process and describes the combined etiology of the disease, with genetic and developmental causes to be responsible for the disorder.[4-6] In some cases when DDH is misdiagnosed, symptoms may appear in adolescence and cause severe degenerative joint disease in early adulthood.6 Different risk factors as family history, breech positioning in utero, being the first-born child, oligohydramnios, female sex, and deformities (postural or structural) of the foot and torticollis have been reported. Historically, Dupuytren described with accuracy this condition based on anatomy, pathology and clinical presentation in 1820s.[7] The diagnosis of DDH was based in clinical examination (Barlow and Ortolani tests), in clinical presentation, radiographs and in 1980, Prof. Reinhard Graf presented his ultrasonographic technique that nowadays dominates as the easiest and most useful screening ultrasound technique.[8] Graf method evaluates the quality of the infantile hip joint, and more specifically, the bony and cartilaginous coverage of the femoral head from the acetabulum by counting specific angles (angle α and angle β).[9] The significance of Graf method is that according to description and these measurements, there is a specific classification (types I to IV) for the DDH that is a guide for the selection of the optimal treatment option. (**Table 1**) Graf method for screening infantile hips has been adopted by the healthcare systems in many European countries as Germany, Austria and Switzerland leading to an important decrease of neglected cases of DDH and to a reduce of open reductions and osteotomies.[10]

Developmental Dysplasia of the Hip (DDH)

The mean incidence of DDH in children without associated risk factors is 11.5/1000 live births according meta-analyses protocols and varies from 0,06/1000 in Africans to 76.1/1000 in Native Americans and Laplanders. DDH appears more frequently in females (19/1000) than males (4.1/1000) and when there is positive family history the risk is 1.7 times higher.[11] DDH is more common in the left hip (64%) than in the right hip (36%) and is usually unilateral (63%).[12] Breech presentation of the infant is connected with high incidence of DDH (7.1% to 40%).[13] Recent methods have proven that feti that are in breech position in utero life have lower incidence of DDH when delivered with Caesarean section, but as expected, higher incidence than children born in vertex position.[14] Family also increases the possibility of DDH presentation.[15] Statistically significant correlations have been found among first-, second-, third-degree relatives and also among siblings, uncles/aunts and cousins.[16,17] Additionally, firstborn children have higher incidence of DDH. Other disorders that are related to DDH are hormonal diseases of the infants (increase in urinary excretion of conjugated estrogen and 17 β-estradiol) and oligohydramnio.[18] Literature is not clear about the role of swaddling, but it seems that populations worldwide that use swaddling for the newborns show higher incidence of DDH.[19,20] Genetics are recently being studied for possible connection with DDH. Until now, different studies have shown correlation between the presence of Human Leucocyte Antigens (HLA A1 and HLA DR4) and DDH and the existence of an autosomal dominant genetic mechanism and a two-gene system of genes (dominant for joint laxity and polygenic for acetabular dysplasia) that is accepted by scientific community.[21,22] Risk factors developed during pregnancy are hypothyroidism or phenylketonuria of the mother, taking progesterone in the 1st trimester for any reason and older parental age.[23-25]

Abnormalities in bones and soft tissues of the hip joint are usual in patients with DDH. The normal

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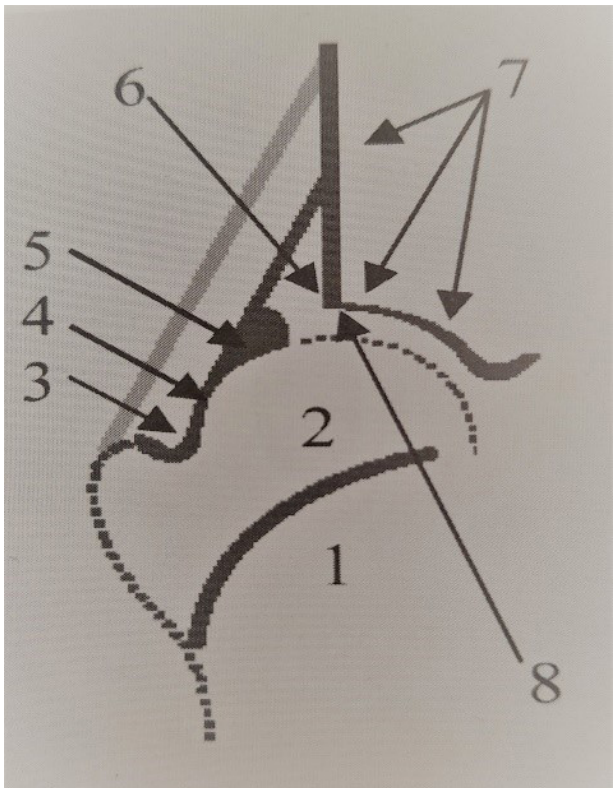

Figure 1: *Infant hip joint anatomical structures: 1. Chondro-osseous border, 2.Femoral head, 3 Synovial fold, 4. Joint capsule, 5. Labrum, 6. Hyaline cartilage, 7. Acetabulum (bone) , 8. Bony rim*

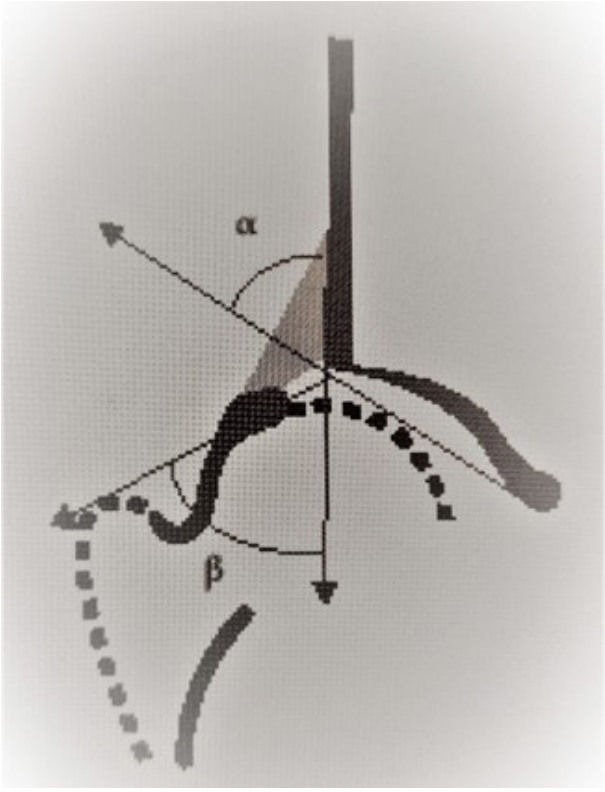

Figure 2: *Angles a and β in Graf ultrasonography method*

hip joint has a ball-socket shape with deep acetabulum in utero life, that becomes shallow at birth and as the newborn develops, becomes deeper and finally covers totally the femoral head. In DDH, the acetabulum remains shallow and the femoral head grows in non-anatomical position. The acetabular index enables to evaluate the coverage of femoral head by acetabulum. The normal range is 3° to 13° and above 13°, there is suspicion of DDH presentation.[26] If the femoral head is positioned out of the acetabulum, the epiphysis develops slowly, the femoral head is flattening and aseptic necrosis is not unusual. In these cases, the labrum of acetabulum becomes hypertrophic, leading to the creation of a secondary "false" acetabulum posteriorly and superiorly of the original one. Even more, a case of double femoral head in DDH patient has been reported as a complication. The femoral neck becomes thick and short and the anteversion of the femoral head changes. As a result, the architecture of the joint is disturbed. Soft tissues of the hip joint also get affected in DDH patients. The articular capsule is thicker in newborns with this disorder and often appears being stuck on the superior and posterior aspect of the acetabulum.[27] The glenoid labrum that is part of acetabular labrum, positioned posterosuperiorly is usually attached at the femoral head and the articular capsule in one side and the cardilaginous part of the roof in the other and along with the hyaline cartilage of the acetabular roof (the epiphyseal plate that deepens the acetabulum) block the reduction of the femoral head.[28,29] It is also described that the acetabulum of DDH patients fills with fat, the empty space is occupied and the closed reduction becomes impossible in neglected cases.[30] The iliopsoas tendon comes in front of the articular capsule, also diminishing the possibility of reduction. When DDH is unilateral, the pelvic inclines and the spinal

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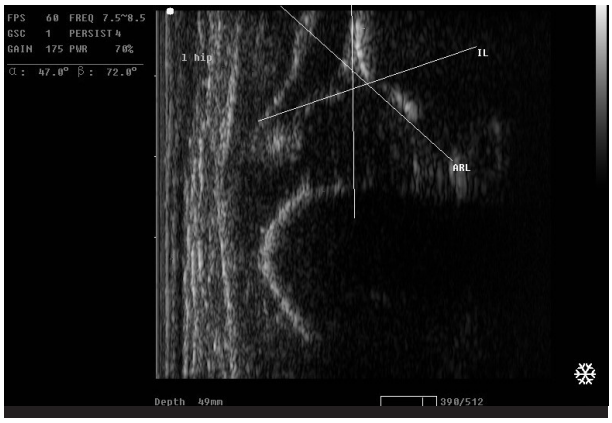

Figure 3: *Sonographic image of infant with a-angle = 47o and β-angle = 72o, type IIc in Graf classification*

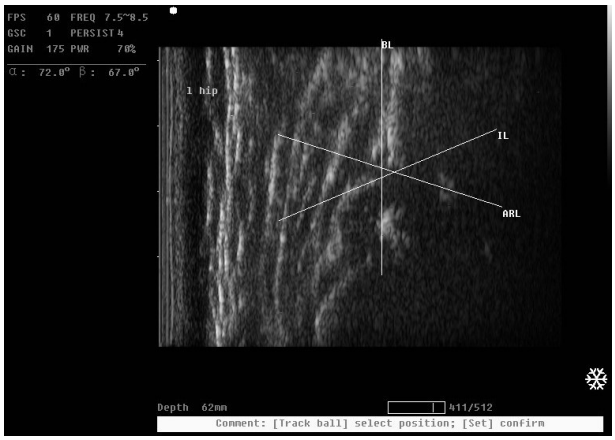

Figure 4: *Sonographic image of infant with a-angle = 72o and β-angle = 67o, type Ib in Graf classification*

curve changes. Furthermore, the abductors of the hip thicken as age of the patient increases, leading to length asymmetry of the lower extremities and hip joint instability.[31] In cases of bilateral DDH the vertical spinal balance changes (increased lumbar lordosis and hip kyphosis), waddling gait is developed.[30] Histologic examination of DDH joints has shown that the articular capsule contains bundles of collagen fibers, thicker than in normal cases, with irregularly distributed fibroblasts internally and also Increased number of elastic fibers, together with chondrocytes of irregular shape. The labrum is inclined to adhere the perichondrium of the outer ilium or the femoral head and shows fibrous metaplasia. The perichondium and the bone around the cartilage is usually normal, but the histology of the acetabulum changes as the growth plates of, ischial, pubic and especially of iliac bones are affected. The ligaments of the hip joint contain randomly arranged collagen bundles and elastic fibers. The vessels of these zones are thick and dilated. The acetabulum becomes anteverted and less vertically inclined and the femoral neck-shaft angle is usually normal or slightly valgus. The delay of growth of femoral head leads to small and spherical shape, or flattened shape in cases of high dislocation. When early closed reduction is successful, the femoral head grows normally.[32]

DDH contains a wide spectrum of hip disorders, from dysplasia (shallow and undeveloped acetabulum) to the teratologic hip.[33] The physical examination is crucial to recognize DDH indications. The most useful tests for the neonatal hip are the Ortolani and Barlow maneuvers. Positive results in these tests are strong indications of luxation of the hip.[34] Furthermore, Galeazzi sign is often used used between 3 and 6 months (with the child in supine position and with the knees and hips in flexion, the examiner notices if the extremities have the same length). Bilateral DDH is more difficult to diagnose without radiological imaging. The median walking age of children with DDH does not differ significantly from healthy children and during growth the patient may develop Trendelenburg sign (opposite side of pelvis dips during one legged stance).[35] Nowadays, DDH is diagnosed with ultrasonographic examination and is usually treated conservatively in infant age. In rare neglected cases, DDH leads to a progressive disorder that badly affects the every-day routine and life of an adolescent and an adult. The physicians must be suspicious for the presentation of the disorder and to suggest hip sonography with Graf method for every newborn as a screening method.

Although physical examination is very important, imaging of the infantile hip is appropriate, not only when there is the suspicion of DDH existence, but for all the newborn infants, because the usual tests (Barlow and Ortolani maneuvers) are not absolutely accurate.[36] Ultrasonograhy is the most useful option for the diagnosis of DDH**.** In 1980, Prof. Graf

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TABLE 1.

Graf classification

Type	Description	Bony Roof	Bony Rim	Cartilage Roof	α- angle (deg.)	β-angle (deg.)	Subtype
I	Mature hip	Good	Angular/ blunt	Covers the femoral head	≥ 60		Ia: β ≤ 55° Ib: β > 55°
IIa	Physiologically immature (< 3 months)	Deficient	Rounded	Covers the femoral head	50-59		IIa+ : α = 55o-59° IIα- : α = 50o-54° (at 6 weeks of age)
IIb	Delay of ossification (> 3 months)	Deficient	Rounded	Covers the femoral head	50-59		
IIc	Critical hip	Severely deficient	Rounded to flattened	Covers the femoral head	43-49	<77	Stable (β < 77°) or unstable (β > 77°) under pressure
D	Decentring hip	Severely deficient	Rounded to flattened	Displaced	43-49	>77	
III	Dislocated hip	Poor	Flattened	Pressed upward, perichondrium slopes cranially	< 43	>77	IIIa: hypoechoic cartilage acetabular roof IIIb: hyperechoic acetabular roof
IV	Dislocated hip	Poor	Flattened	Pressed downward, perichondrium dips caudally	< 43		

presented his technique to study and evaluate infantile hips for the possibility of DDH presentation. He suggested that ultrasonography study should be included as screening test in all newborns, regardless of any risk factor presence. Nowadays, hip sonography with the Graf method is included in mandatory screening examination of the newborns in Germany, Austria, Switzerland, Czech Republic, and in other European countries.[37] Additionally, other sonographic techniques have also been developed (e.g., the Harcke, Terjesen, and Suzuki methods), but the Graf method has dominated because of its standarized examination technique and of its high sensitivity and specificity.[38] Anatomy of the infantile hip is important for the Graf technique. The distal femur consists of hyaline cartilage (femoral head part of the neck and the trochanter) and between the cartilage and the osseous parts, there is the chondroosseous border, which guides the identification of the other anatomical structures during hip sonography. The femoral head has oval shape and the nucleus (ossification center) is seen in ultrasonographic examination 4–8 weeks earlier than in radiographs. The synovial fold penetrates the femoral neck .The joint capsule includes the femoral head with cranial direction to the rectus femoris muscle. Medially of the capsule is located the labrum of the acetabulum, which has triangular shape and is connected to the acetabular cartilaginous roof and is needed to be identified in order to have a proper ultrasound examination. Finally, the bony rim of the acetabulum is a very significant anatomical structure that distinguishes the osseous and cartilaginous parts of the acetabulum and gives us information about the coverage of the femoral head by osseous acetabulum. In ultrasonographic examination it is the spot where concavity switches to convexity in acetabulum.[39] In order to have accurate ultrasonographic examination, the physician must recognize all the following anatomical struc-

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tures (**Fig. 1**): chondrooseous border, femoral head, synovial fold, joint capsule, labrum, hyaline cartilage, acetabulum (bone), and bony rim (concavity to convexity). In the evolution of the human species, the posterior osseous part of the acetabulum is more developed than the other parts because of the bipedal gait and the standing position, but the middle part is the more significant, because is the location where the load is transported from the corps to the lower limbs. For this reason, we select sections of the middle acetabulum to evaluate the joint in hip sonography of infant hip.[40] Radiography is useful for the study of DDH at the age of 4–6 months or more, when the ossification center of the femoral head appears and can be recorded in radiograph. Anteroposterior and frog position radiographs are to show possible reduction. In radiographs, the most useful signs are the Shenton line and the acetabular index. Radiography is also useful for the follow-up of infants to be treated with Frejka pillow and Pavlik harness and for the intraoperative imaging of the reduction when hip spica is put.[41] CT scan and MRI are widely used for the diagnosis of DDH. CT is mainly in adolescent and young adults as part of preoperative plan, before operations including pelvic and femoral osteotomies. MRI is impossible to be performed for neonates because it requires 30 minutes of isolation in the machine and is rarely used in adolescent and young adults to note labral abnormalities.[42]

The Graf Method

Graf method for the diagnosis of DDH in infants is the gold standard of hip sonography and has dominated on other ultrasonographic techniques because of its high sensitivity, specificity and relationship with a specific classification, which guides the physician to select the optimal treatment option.[43] Physicians who perform ultrasonographic examination should have deep knowledge of the anatomic area. As already, mentioned, for an accurate examination the examiner has to be able to see and recognize all the following structures: chondrooseous border, femoral head, synovial fold, joint capsule, labrum, hyaline cartilage, acetabular bony roof, bonny rim (concavity to convexity). In ultrasonographic examination with Graf method for the infant hip, the physician has to be able to find and signal the exact spot where concavity switches to convexity in acetabulum.[39] The second part of sonographic examination is the usability check. More specifically, during the examination the hip joint has to be depicted in a standardized plane. The lower limb of iliac bone is more or less the center of the acetabulum, next to the triradiate cartilage and it has to be visible during the examination to make it valid.[44] The examiner puts the ultrasound transducer in the coronal plane and moves backwards and forwards to illustrate the lower limb as big and clear as possible. Then he freezes the image and checks the plane. He rotates the transducer appropriately and repeats the forward-backwards movement. This process is repeated until lower limb and correct plane are both visible. For the accurate evaluation and classification of the infant hip joint with sonographic examination, Prof. Reinhard Graf described angles α an β, that are measured through the examination and give specific information if the joint is normal or if there is DDH. The examiner designs the bony roof line, a line that passes through the lower limb of ilium in contact to the lateral limit of osseous acetabulum and also the base line, a line that connects the point where the proximal perichondrium meets the iliac bone and is in contact to the outer border of the pelvic bone.[9] The angle that is formed between these two lines is the α angle and signals the osseous coverage of the femoral head by the acetabulum. Another line that begins from the bony rim and follows a straight direction to the labrum is called cartilage roof line and together, with the base line they include an angle (β angle), which defines the cartilaginous coverage of the femoral head by the acetabulum. Being able to measure these two lines gives the examiner the ability to classify the hip joint via Graf method and classification. (**Fig. 2**) The mother also takes part in the examination process, being present during the whole procedure and is responsible to keep the infant calm. The infant is put by the examiner in a specialized device (Sono-Fix) in lateral position with the right hip up. The examiner goes posteriorly of the infant, using the ultrasound machine (Sono-Guide), examining

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both hip joints by measuring angles α and β. The mother stays in the frontal side, calming the infant. Sometimes, after the ultrasonographic examination, physical examination with Barlow and Ortolani maneuvers is still used for the evaluation of the newborn.[45]

The measurement of α and β angles enables the physicians to classify easily the infantile hips and proceed to the optimal treatment. Graf classification has 4 types and each type is treated with specific option (no treatment, follow-up or medical intervention with conservative treatment). In Type I belong all infant hip joints with the following angle measurements: angle α 60° or more. When angle β is <55° , the subtype is Iα and when angle β is 55° or more, the subtype is Iβ. Both subtypes are normal and no treatment is needed. In Type II belong cases with angle α between 43°-59°. There are subtypes IIα (IIα+ and IIα-) , IIβ, IIc and D. Type IIα are the hip joints with angle α between 50°-59° in babies younger than 3 months of age. Type IIα+ describes immature joints that will probably grow normally the first 3 months (that means with an α angle more than 55° after 6 weeks) and is also a normal condition. Cases that are classified as Type IIα-, are not expected to develop normally in the first 3 months (α angle less than 55° after 6 weeks) and the hip joint needs specific treatment. In type IIβ the angle α is between 50°-59° in child older than 3 months and the joint is defined as dysplastic and also requires specific treatment. Type IIc hip joints with angle α between 43°-49° are considered as dysplastic and immediate treatment is needed. When angle β is bigger than 77° and angle α is between 43°-49°, the joint is considered as type D in Graf classification the femoral head is decentering (dislocating).[46] Finally, types III and IV include cases with angle α less than 43° . In these cases, the infants have decentered (dislocated) hips and immediate treatment is appropriate. In type III, the femoral head pusses upwards the cartilaginous roof and in contrast, in type IV hip joints, the femoral head is completely dislocated posteriorly and upwards and pushes the cartilaginous roof down.[47] (**Fig. 3,4**) Hip sonography with Graf method enables physicians to evaluate properly infantile hip joints, to classify DDH cases with accuracy and to begin with specific treatment when is required. Graf method requires a well trained and experienced physician, because mistakes in angles measurement lead to false results and to non-suitable treatment selection. The main mistakes during hip sonography that may lead to inaccurate measurements are wrong anatomical identification of the structures of the joint and tilting effects due to incorrect positioning of the infant.[48]

Discussion

Musculoskeletal ultrasonography is very useful for soft tissue evaluation and is also widely used to find fluid collection inside the muscle mass or in the joints, or to visualize cartilage and bone surface. With dynamic sonographic examination, examiners are able to assess tendon movement, ligamentous injury, nerve compression and joint.[49] Preventive ultrasound examination in infant age for the diagnosis of DDH has been widely worldwide. Graf method is the most popular among ultrasonographic methods for the diagnosis of DDH and with Graf classification, the responsible physicians are able to categorize the infantile hip joint and to proceed to the appropriate therapeutic protocol, if needed. In cases, when DDH is diagnosed and re-examination or therapeutic intervention is needed, informing the parents is very in order to prepare for the possible long-term treatment and compliance required to achieve centering of the hip joint without the need for surgery.[50] Hip sonography as screening method for the presence of DDH has been established as a standard process in many European countries. The main reason that hip sonography with Graf method should be mandatory screening test for all newborns isa that is a very accurate method that reduces the cases of neglected DDH who develop dysplastic hip joints in early adulthood and to reduce the complex pelvic operations in children with diagnosed DDH, by using the proper conservative treatment option set by Graf classification.[51] It is proven in literature that after the establishment of Graf method as standard screening method for DDH, the open reductions and acetabuloplasties have decreased dramatically.[52] It also seems that Graf method is

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cost-effective as it is much cheaper than the complex operations and rehabilitation programs that were required before the establishment of national screening programs.

The period of time between 4th and 6th week of life is the best for the hip sonography and it is important that the examiner is certified for the Graf method, because possible failures in the process cause errors in classification errors in treatment. Usually, the examiner is orthopedic surgeon or radiologist and is preferable that the examiner is also responsible for the treatment procedure.[53] Another important fact for the Graf method is that this technique is characterized by repeatability. Different studies have shown that experienced examiners have made the same measurements in the same infant hips and the same classification by reading the same ultrasound images, making the method suitable for interdisciplinary communication. Graf method is the most used sonographic technique for the screening of DDH in infants because of the correlation of the measurements (angles α and β) with the classification and therapy. The results have been investigated in literature and it has been proven that since the decade of 1980, when ultrasonographic examination of the hip have commenced (Graf 1980, Harcke 1984, Suzuki 1987 and Terjesen 1988), the neglected cases of DDH have decreased significantly, decreasing the number of adolescents and young adults with dysplastic hip joints.[54] Every newborn and not only when there are known predisposing factors (female gender, positive family history, breech position in utero, low birth weight, oligohydramnios etc) or suspicion during the clinical examination (positive Barlow and Ortolani tests)should undergo hip sonography as screening test.[55,56] Supposing that the screening was selective only to infants who present one or more of the risk factors, the number of the sonographic examinations would also be large, because in essence, only boys without any predisposing factor would be excluded from the examination. Furthermore, published studies form Germany and Austria, where the infant hip screening with Graf method is universal , have shown that after the establishment of this examination as mandatory screening test, very few open reductions have been carried out.[57,58]

According to sonographic findings, Graf method is more effective for children aged form 28-77 days. The best period for the sonographic examination is between the 4th and the 6th week, a period when the hip joint is demarcated well and the doctor is able to commence the appropriate treatment protocol if needed.[59] Graf method seems to be the most reliable method for the screening of DDH as it gives clear information for the pathology of the infantile hip joint and simultaneously enables the examiner to classify the examined joint in a strict classification. A study from Japan showed that the physicians who were trained in just one course for Graf method had a small learning curve and after the course fell safe and confident to perform hip sonography.[60] More orthopaedic surgeons should learn Graf method, because it allows them to understand a DDH case in very early and enables proper conservative treatment without needing complex pelvic or femoral osteotomies.

Outcome

Developmental Dysplasia of the Hip (DDH) is one of the most common musculoskeletal disorders in infants. In previous years the majority of DDH patients were unsuccessfully treated, with treatment protocols that required open reductions and there were also cases that were misdiagnosed and patients developed osteoarthritis of the hip joint in adolescence. The development of ultrasonographic techniques enabled the orthopedic surgeons to diagnose DDH in early stage and to perform the suitable treatment in time. Through the years, different sonographic techniques have been developed, static or dynamic which contributed to the reduce of neglected DDH cases. The most dominant sonographic technique for the diagnosis of DDH is Graf method. Graf method evaluates the osseous and the cartilaginous coverage of the femoral head by the acetabulum, measuring angles α and β respectively. This specific method has objective measurements that are based on anatomical correlations and correspond to a certain classification. Four different types (I, II, III, IV) and some subtypes are included in Graf classification for the DDH and each of them match-

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es with a specific treatment option. In many countries of Central Europe Graf sonographic method for the diagnosis of DDH is included in the mandatory preventive check-up for the infants in the first days after birth. It is proposed that ultrasonographic examination is appropriate only in infants with risk factors for DDH, but in this way no standarised guidelines can control for which children have to be examined. The preventive sonographic examination has increased the accuracy of finding DDH cases in relation to simple clinical examination (Barlow and Ortolani tests) and decreased the neglected DDH cases that required surgical treatment with major operations and pelvic osteotomies. As it has been proven that Graf method is the most accurate and useful, it has to spread furthermore. More orthopaedics should be trained in the this technique in order to be able to evaluate the infantile hip joints and to perform treatment where needed. Hip sonography with Graf method made DDH an easily cured disease treated with conservative means with great success, without requiring complex pelvic operations for the children or total hip arthroplasties in early age during adolescence or adulthood.

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