

# Developmental Dysplasia of the Hip: A Review

Spyridon Sioutis,<sup>a</sup> Stylianos Kolovos,<sup>b</sup> Maria-Eleni Papakonstantinou,<sup>c</sup> Lampros Reppas,<sup>a</sup> Dimitrios Koulalis,<sup>a</sup> & Andreas F. Mavrogenis<sup>a,\*</sup>

<sup>a</sup>First Department of Orthopaedics, National and Kapodistrian University of Athens, School of Medicine, Athens, Greece; <sup>b</sup>Department of Orthopaedics, General Hospital of Larisa, Larisa, Greece; <sup>c</sup>Department of Pediatrics, General Hospital of Kefalonia, Kefalonia, Greece

\*Address all correspondence to: Andreas F. Mavrogenis, MD, First Department of Orthopaedics, National and Kapodistrian University of Athens, School of Medicine, 41 Ventouri Street, 15562, Holargos, Athens, Greece; Tel./Fax: +0030-210-6542800, E-mail: afm@otenet.gr

**ABSTRACT:** Developmental dysplasia of the hip (DDH) is the most common musculoskeletal disorder of the infant age. Its incidence ranges from 0.06/1000 to 76.1/1000 live births and is more frequent in female infants. Breech position, family history and firstborn children are the main risk factors for DDH and this disorder is also associated with the presence of other congenital deformities. Anatomically, the acetabulum remains shallow and the femoral head grows in a wrong position. Clinical examination is important and tests such as Barlow and Ortolani give indications only for a part of the spectrum of this entity. Nowadays the sonographic examination is the most accurate option for the diagnosis. Graf classification categorizes the DDH cases in four types, from normal to dislocated hip, by description and measuring specific angles in sonographic examination. The wide usage of ultrasonography has decreased the non-diagnosed or neglected cases; treatment begins immediately in young age and is usually conservative with the usage of devices such as Pavlik harness and hip spica. To enhance the literature, we searched for published studies on DDH, to summarize the pathogenesis and the diagnosis and to discuss the treatment and outcome of the patients with this disorder.

**KEY WORDS:** DDH, developmental dysplasia of the hip, hip ultrasonography, Graf classification, Pavlik harness, hip spica

## I. INTRODUCTION

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.<sup>1</sup> Formerly known as congenital dysplasia of the hip, DDH not only defines congenital malformation, but also includes perturbations during development process.<sup>2</sup> The term of developmental dysplasia of the hip describes the combined etiology of the disease, with genetic and developmental causes to be responsible for the disorder.<sup>3,4</sup> Snyder et al. first used the term “developmental dysplasia of the hip” in 1992.<sup>5</sup> In some cases when DDH is misdiagnosed, symptoms may appear in adolescence (late dysplasia), and it is confirmed that untreated dysplasia leads to degenerative joint disease.<sup>6</sup>

Historically, the possibility of congenital hip in newborns has been known since the days of

Hippocrates and in medieval London this entity was common.<sup>7,8</sup> Paletta, a Milanese physician, and Dupuytren described with accuracy this condition based on anatomy, pathology and clinical presentation in 1820s.<sup>9,10</sup> The diagnosis of DDH was based in clinical examination (Barlow and Ortolani tests), in clinical presentation, radiographs and it was only in 1960s that ultrasound screening has been widely used in developed countries.<sup>11</sup> In 1980, Prof. Reinhard Graf presented his ultrasonographic technique that nowadays dominates as the easiest and most useful screening ultrasound technique.<sup>12</sup>

As for the treatment options, early reduction and treatment techniques were not effective and it was Lorenz in 1986 that proposed closed reduction and immobilization with a plaster cast achieving successful reduction in full abducted position but often producing femoral head necrosis.<sup>13</sup> In the 20th century more sophisticated means of in-plaster positioning and maintaining devices as Pavlik harness and Frejka pillow have been developed for mild severity cases and Salter and other osteotomies have

been described for neglected cases or when conservative treatment fails.<sup>14</sup>

## II. EPIDEMIOLOGY AND RISK FACTORS

The mean incidence of DDH in children without associated risk factors is 11.5/1000 live births according to meta-analyses protocols and varies from 0.06/1000 in Africans to 76.1/1000 in Native Americans and Laplanders.<sup>15,16</sup> In the United Kingdom, the incidence is 5/1000.<sup>17</sup> The disorder is more frequent in females (19/1000) than males (4.1/1000) and the relative risk with positive family history is 1.7 times higher.<sup>15</sup> Because of the prevalence of the disorder in newborn females, the majority of cases presented in literature concern the clinical presentation in girls. According to Goiano et al., male patients present different characteristics as higher frequency of bilateral disease, later diagnosis and a higher percentage of high dislocations than in other studies.<sup>18</sup> DDH is more common in the left hip (64%) than in the right hip (36%) and is usually unilateral (63%).<sup>15,19</sup>

Breech presentation is connected with higher incidence of DDH and ranges from 7.1% to 40% for neonates born in this position.<sup>20–23</sup> The type of breech position (frank, incomplete, complete) is also important for the presentation of and the frank breech position seems to be a higher risk factor.<sup>24</sup> According to recent studies, newborns in breech position have lower incidence of DDH when delivered with Caesarean section than in normal vaginal delivery, but as expected, higher incidence than children born in vertex position.<sup>25,26</sup>

Family history is another risk factor that increases the possibility of DDH presentation.<sup>27,28</sup> In recent studies has been a correlation of first-, second-, and third-degree relatives.<sup>29,30</sup> Especially in Japan, the Middle East, Mediterranean countries, and in Native Americans, 14% to 49% of children born with DDH have one of their parents with the same disorder.<sup>27,31–33</sup> Similar correlations have been found among siblings, uncles/aunts and cousins.<sup>34</sup>

Firstborn children have higher incidence of DDH. Evidence does not approve correlation between twins, fact that occurs that the specific disorder is supported by a polygenic mechanism.<sup>35–37</sup> In contrast, the incidence of the disease decreases

in premature and low-birth-weight infants, even in children born in breech position.<sup>24,38</sup> Other conditions that are related to DDH are hormonal disorders of the newborn (increase in urinary excretion of conjugated estrogen and 17  $\beta$ -estradiol), pelvic joint instability and pain of the mother during pregnancy and oligohydramnio.<sup>39–41</sup>

The literature is not clear about the role of swaddling. There is evidence based studies for Native Americans that show no statistically significant difference for DDH presentation in newborns carried in cradleboards. Similar results are presented in studies from Iran and Saudi Arabia, where swaddling is also usual.<sup>27,42,43</sup> In most studies, a strong correlation between swaddling and DDH has been proved. Studies for Canadian Native Americans, and studies from Turkey and Hungary, where this technique for carrying babies is usual, show that there is a higher incidence of DDH.<sup>44–46</sup> Two interesting facts prove that this correlation probably exists: the reduction of cases of DDH in Qatar community after a program that informed the locals for the harmful consequences of swaddling from 20% to 6%; and that the two main Arctic populations (Sami and Inuit/Eskimos) have remarkably different incidences of the disease. The first group used swaddling in the past and has high incidence of hip dysplasia, while Eskimos mothers carry their babies on their backs with the hips abducted and have similar incidence of DDH with Caucasians.<sup>47–49</sup> Furthermore, in places where swaddling is not in their culture as Thailand, China and African countries the incidence is low.<sup>50,51</sup> As for seasonal variation, most studies show higher incidence of the disorder in children born in autumn and winter and this probably happens because of the extended usage of swaddling and similar clothes in cold months.<sup>29,32,52</sup>

Genetics are being studied for their role in DDH. Earlier studies showed the existence of an autosomal dominant genetic mechanism in France and Turkey and a two-gene system of genes (dominant for joint laxity and polygenic for acetabular dysplasia) is accepted by scientific community.<sup>30,53,54</sup> Studies in Greece and Japan show correlation between the presence of Human Leucocyte Antigens (HLA A1 and HLA DR4) and DDH.<sup>55,56</sup> There is not significant evidence about gene mutations to be

associated with hip dysplasia except of 4q35 mutation (single family in South Africa) and 17q21 mutation (China).<sup>57,58</sup>

During pregnancy, many factors may affect the risk of DDH presentation. At first, if mother has hypothyroidism or phenylketonuria, the incidence of the disease increases.<sup>59,60</sup> Taking progesterone in the 1<sup>st</sup> trimester for any reason, also increases the chance of DDH appearance.<sup>61</sup> According to Chasiotis-Tourikis et al., smoking during pregnancy may decrease the risk in female newborns, but not in male.<sup>62</sup> Older parental age seems to increase the chance of DDH presentation.<sup>63</sup> Studies have proved that viruses and other microbes do not play a role in the incidence of the disorder.<sup>64,65</sup> Amniocentesis during the first trimester likely decreases the possibility, while during the second trimester does not change the risk.<sup>66,67</sup> The socioeconomic level of the family does not play any role in the incidence of the disorder in the majority of studies.<sup>26,68</sup>

DDH is associated with congenital deformities such as congenital muscular torticollis, congenital foot deformities such as clubfoot or metatarsus adductus, infantile scoliosis, and other spinal disorders such as spina bifida occulta.<sup>69–74</sup> Finally, studies have proved that hip muscles in the diseased hip become atrophic and children often develop pelvic asymmetry.<sup>75–78</sup>

### III. PATHOPHYSIOLOGY

Abnormalities in bones and soft tissues of the hip joint are usual in patients with DDH. The skeletal changes affect on the acetabulum, on the femoral head and neck and on the pelvis due to subluxation or dislocation.<sup>79</sup> Normally, the 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.<sup>80</sup> In DDH presentation, the acetabulum remains shallow and the femoral head grows in a wrong, non-anatomical position. The best way to evaluate the coverage of femoral head by acetabulum is the acetabular index, an angle that can be measured in anteroposterior radiograph by a horizontal line and a line drawn running through the medial edge of the sclerotic acetabular zone and

through the lateral sourcil. The normal range is 3° to 13° and when the angle is above 13°, there is suspicion of DDH.<sup>81</sup> In addition, the acetabulum thickens and acquires increased anteversion.<sup>82</sup>

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.<sup>83</sup> 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.<sup>84,85</sup> Even more, a case of double femoral head in DDH patient has been reported as a complication.<sup>86</sup> The femoral neck becomes thick and short and the antversion of the femoral head changes. As a result, the architecture of the joint is disturbed.<sup>87</sup>

The 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.<sup>84,88</sup> 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.<sup>84,89,90</sup> The ligaments become elongated and hypertrophied in most cases, fact that also complicates the reduction.<sup>1</sup> It is also described that the acetabulum of DDH patients fills with fat, the empty space is occupied and the closed reduction becomes impossible.<sup>79</sup> Additionally, the iliopsoas tendon may come in front of the articular capsule, increasing in this way the problem of stenosis and also diminishing the possibility of reduction.<sup>1,91</sup>

In unilateral DDH, the pelvic inclines and the spinal curve changes. Furthermore, the abductors of the hip and especially the gluteus medius lose part of their volume and strength as age of the patient increases, leading to length asymmetry of the lower extremities and joint instability.<sup>92</sup> In cases of bilateral DDH the vertical spinal balance changes (increased lumbar lordosis and hip kyphosis) and if the patient is not treated, waddling gait is developed.<sup>79</sup>

As for histology, the articular capsule contains bundles of collagen fibers, thicker than in normal

cases, with irregularly distributed fibroblasts internally. Increased number of elastic fibers, together with chondrocytes of irregular shape are also comprehended in the tissue.<sup>93</sup> The labrum is inclined to adhere the perichondrium of the outer ilium or the femoral head and shows fibrous metaplasia. The perichondrium and the bone around the cartilage is usually normal.<sup>94</sup> The ligaments of the hip joint contain randomly arranged collagen bundles and elastic fibers.<sup>93</sup> The histology of the acetabulum changes dramatically, as the growth plates of, ischial, pubic and especially of iliac bones are affected. Irregularly arranged chondrocytes appear with a tendency to lead to degenerative lesions. The vessels of these zones are thick and dilated and temporary partial microepiphysiodesis is usual and corrupts the acetabulum architecture in relation to the pelvis. During developmental process, the acetabulum becomes anteverted and less vertically inclined.<sup>1,95</sup> The femoral neck-shaft angle is usually normal or slightly valgus (normal range  $134^{\circ} \pm 10.8^{\circ}$ ) and the anteversion depends on the position of the dislocated femoral head. The growth of femoral head delays, leading to small and spherical shape, but the shape of femoral head is flattened in cases of high dislocation. When early closed reduction is successful, the femoral head grows normally. Growth plate destruction during the reduction process results in malformation (trochanteric overgrowth and a short and varus femoral neck). Histologically, the findings in the femoral head are similar to the acetabulum, with presence of chondrocytes and areas of calcification.<sup>88,95-97</sup>

#### IV. CLINICAL PRESENTATION AND PHYSICAL EXAMINATION

DDH contains a wide spectrum of hip disorders, from dysplasia (shallow and undeveloped acetabulum) to more severe dysplasia (displacement of the joint, but the connection of articular surfaces remains stable) and severe dysplasia-dislocation (displacement, without articular surfaces connection) and the most severe form, the teratologic hip.<sup>98</sup> Teratologic hip is an entity, in which the hip is in utero dislocated and usually irreducible when the child is born. It is commonly associated with neuromuscular

conditions and other disorders as myelomeningocele, arthrogryposis and Ehlers-Danlos disease.<sup>99</sup>

The physical examination of the newborn is very important to recognize DDH indications. The most useful tests for the neonatal hip are the Ortolani maneuver, in which a “clunk” is felt when the femoral head is moved in the acetabulum and Barlow maneuver, in which the physician dislocates the hip and a “clunk” is also felt.<sup>100,101</sup> In the Ortolani maneuver, the hip is gently abducted with knees and hips in  $90^{\circ}$  flexion. Positive results in Barlow and Ortolani tests are strong indications of luxation of the hip.

One more useful sign that can be used between 3 and 6 months is the Galeazzi sign. Probably, at this age DDH will have been recognized from ultrasound screening test and the infant is under therapy. In cases of dislocated hips that are undiagnosed, Galeazzi sign gives indications of the disorder. In particular, with the child in supine position and with the knees and hips in flexion, the examiner notices if the extremities have the same length. If any of them is shorter, DDH is the most common diagnosis, but further investigation with ultrasound or radiographs is necessary.<sup>102</sup> Additionally, other signs of late diagnosis are decreased abduction of affected hip, asymmetry in gluteus strength. Bilateral DDH is more difficult to diagnose without radiological imaging and clinical suspicions are waddling gait and hyperlordosis.<sup>103</sup>

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).<sup>104,105</sup> When DDH is not well treated in infant age, it progresses in childhood dysplasia and later on in adult dysplasia. In these cases, studies show that these patients have annoying symptoms in everyday life and undergo a total hip arthroplasty operation in young age.<sup>106</sup> It is easily understandable that not only the diagnosis and the proper treatment in infant age is significant, but also the follow-up, because the growth process is a dynamic condition and many changes can happen. There are also asymptomatic cases, usually persons with underlying joint laxity, without symptoms in childhood that



become symptomatic later (asymptomatic radiographic dysplasia).<sup>107</sup>

Adult hip dysplasia is an entity often caused by acetabular dysplasia. The diagnosis is given radiographically by measuring the angle of Wiberg for the lateral coverage of femoral head by acetabulum. In these cases, the patients were asymptomatic in childhood.<sup>108</sup> Another type, the most usual, degenerative disorder of adult hip is osteoarthritis (OA) and can be primary or secondary. In primary OA, the causes are unknown and the diagnosis is getting by exclusion.<sup>109</sup> Secondary OA is caused by already existing disorders of the childhood as DDH, Legg-Calve-Perthes disease and slipped capital femoral epiphysis. As it is referred to the literature, about 20% of hip OA is caused by previous hip dysplasia and the other 80% is due to degenerative articular cartilage or bone lesions.<sup>109,110</sup> OA can be measured either with pelvic radiographs in a population and examine the rate of radiological signs of OA in the amount of this population, or by recording the rates of total hip arthroplasties in a population, with the restriction to include only symptomatic cases of OA.<sup>109</sup>

Today, DDH can be easily diagnosed with ultrasound examination and treated conservatively in infant age. If for any reason it is not treated, it becomes a progressive disorder that badly affects the every-day routine and life of an adolescent and an adult. The physicians must be suspicious for this insidious entity in any indication during clinical examination, because the degenerative lesions caused by the disorder will lead the patient to undergo a major hip surgery in a young age.<sup>106,111</sup>

## V. DIAGNOSIS

DDH is the most common musculoskeletal abnormality in infants and for this reason, diagnosis is very important. Physical examination is the first step of the diagnostic process and all newborn infants should be examined before exiting the nursery. Radiographs were important in previous years for the diagnosis and usage of computed tomography (CT) scan and magnetic resonance imaging (MRI) is rare. In our era, the gold standard of DDH diagnosis is the ultrasonography and more specifically, according to

Prof. Graf, the sonographic study of infant hip must be used as a typical screening test.

As mentioned above, physical examination of the newborn infant is one of the most important stages of the evaluation. Barlow and Ortolani maneuvers are necessary examinations and positive results are strong indications of the disease. In addition, asymmetrical gluteal skin folds, different limp height (supine position with hips and knees flexed) and abduction and adduction maneuvers are included in a comprehensive physical examination of the infant.<sup>112</sup> As the newborn grows, Barlow and Ortolani are getting negative and no specific for possibility of DDH after the age of 3 months. The range of abduction is the most reliable sign by this age until the child starts to walk. After starting to walk, a child with DDH may present with Trendelenburg gait sign. In bilateral disease, children may also present waddling gait and symmetrical, but decreased abduction. Although physical examination is very important, further investigation is needed, not only when there is the suspicion of DDH existence, but for all the newborn infants, because the typical tests are not absolutely accurate.<sup>104</sup>

In 1980, Prof. Graf presented his technique to study and evaluate hips of infants. He also suggested that ultrasonography study should be included as screening test in all newborns and in the next years, this examination became very popular in countries of Central Europe.<sup>12</sup> Nowadays, hip ultrasonographic examination is included in mandatory screening examination of the newborns in Germany, Austria, Switzerland, Czech Republic, and in other European countries.<sup>113</sup> Other sonographic techniques have also been developed (e.g., the Härke, Terjesen, and Suzuki methods), but the Graf method has dominated because of its standardised examination technique and of its high sensitivity and specificity.<sup>114–117</sup>

Anatomy of the hip in infant age is important to understand the usage of Graf technique.<sup>12</sup> 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, an important sonographic structure for the identification of the other anatomical structures. The femoral head has oval shape and the nucleus (ossification center) is seen in

ultrasound examination 4–8 weeks earlier than in radiographs. The synovial fold penetrates the femoral neck with direction to the trochanter and is depicted as a circle in ultrasound. 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.<sup>118,119</sup>

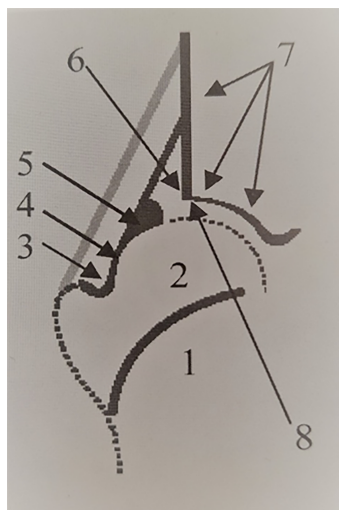
Practically, to have a proper examination, the physician must recognize all the following anatomical structures (Fig. 1): chondroosseous border, femoral head, synovial fold, joint capsule, labrum, hyaline cartilage, acetabulum (bone), and bony rim (concavity to convexity).<sup>12,89,120</sup> The usability check is the second essential step for the evaluation of the ultrasonographic pictures. The examination must be in a standardized plane. In Graf technique, the lower

limb of iliac bone has to be depicted in all images to make a valid examination. The lower limb of iliac bone is the center of the acetabulum, next to the tri-radiate cartilage. In any other case the examination is invalid, except for cases of posterior and cranial dislocation of the hip.<sup>121</sup>

Additionally, in the evolution of the human species, the posterior osseous part of the acetabulum is more developed than the middle or the anterior and because of the bipedal gait and the standing position, the middle part of the acetabulum is the more significant and basic because there 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 ultrasonography of infant hip.<sup>122,123</sup>

During the process, the physician must be careful in every stage. At first, the full documents of the infant should be recorded. The role of the mother is crucial, because she has to be present in order to keep the infant calm. The examiner takes the baby and puts it in the specialized positioning device (Sono-Fix) in lateral position with the right hip up. The examiner goes posteriorly of the infant and the mother in the frontal side and touches it with her right hand in the right shoulder. In the next step, the examiner uses the the specialized ultrasound machine (Sono-Guide) and in vertical capture examines the right and then the left hip. He selects the two best images from each side to measure the  $\alpha$  and  $\beta$  angles and evaluate the hips. When the sonographic examination is finished, the physician should go on with the physical examination using the proper maneuvers and tests.<sup>124,125</sup>

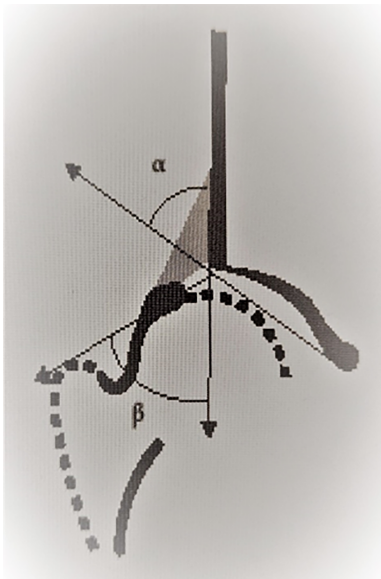
The measurement of  $\alpha$  and  $\beta$  angles is the last stage of ultrasonographic study. Firstly, the examiner designs the bony roof line, a line that connects the lower limb of ilium to the to the lateral limit of osseous acetabulum and then the base line, a line that connects the point where the proximal perichondrium meets the iliac bone and is in contact to the outer border of the pelvic bone.<sup>12</sup> The angle included between these two lines is the  $\alpha$  angle and quantifies the bony coverage of femoral head by the acetabulum.<sup>12,125</sup> Additionally, there is one more line that is scheduled between the bony rim spot and moves forward to the labrum. This line is



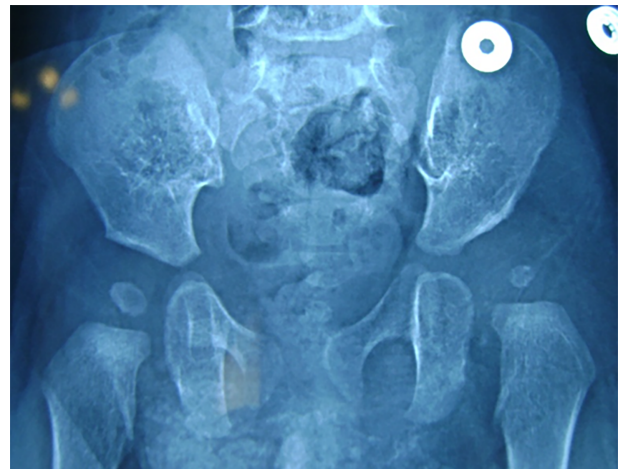
**FIG. 1:** Infant hip joint anatomical structures. 1: Chondroosseous border; 2: femoral head; 3: synovial fold; 4: joint capsule; 5: labrum; 6: hyaline cartilage; 7: acetabulum (bone), 8: bony rim.

called cartilage roof line. The angle that is included between cartilage roof line and base line is the  $\beta$  and defines the cartilaginous coverage of the femoral head by the acetabulum. Respecting the local anatomy, recognizing all the needed structures and having the infant in the right position, the measurement of these angles leads to the classification of the infant hip and the possible selection of proper kind of treatment if needed (Fig. 2).<sup>126–128</sup>

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 (Fig. 3).<sup>129</sup> Anteroposterior radiograph is needed and only if dislocation is recorded, radiograph in frog position is useful to show possible reduction. In radiographs, the most useful signs are the Shenton line and the acetabular index. The Shenton line is an imaginary curved line drawn along the inferior border of the superior pubic ramus and along the inferomedial border of the neck of femur.<sup>130</sup> This line is normally continuous and smooth and is interrupted in DDH in infants or in fractured femoral neck in adults.<sup>128</sup> The acetabular index reports the coverage of femoral head by acetabulum.<sup>81</sup> Radiography is also useful for the follow-up of infants to be treated with Frejka pillow and Pavlik



**FIG. 2:** Angles  $\alpha$  and  $\beta$  in the Graf ultrasonography method



**FIG. 3:** Anteroposterior radiograph of a 6-month-old baby with a dysplastic left hip joint

harness and for the intraoperative imaging of the reduction when hip spica is put.<sup>131,132</sup>

CT scan and MRI are not often used in DDH examination. More specifically, CT scan can be used for assessment of closed or open reduction and mainly in adolescent and young adults as part of preoperative plan, before operations including pelvic and femoral osteotomies. MRI is very difficult to be done for neonates because it requires 30 minutes of isolation in the machine and is used in adolescent and young adults to identify labral abnormalities. Both of these examinations are very rarely used in typical clinical assessment.<sup>133–135</sup>

## VI. CLASSIFICATIONS

The determination of sonographic types is based on lesions in acetabulum of the hip joint and in the range of displacement of the femoral head when dislocated. Therefore, the description of types is a classification of bony and cartilaginous acetabulum in relationship with the growing age of the infant. There are four types (from I to IV) that describe the femoral head coverage (Table 1).<sup>136,137</sup> In type I, the bony roof is well shaped, the bony rim is angular and the cartilage roof covers the femoral head. In type II, the bony roof is deficient, the bony rim is rounded and the cartilage roof also covers the femoral head. Type III describes a joint with poor bony

**TABLE 1:** Graf classification of DDH

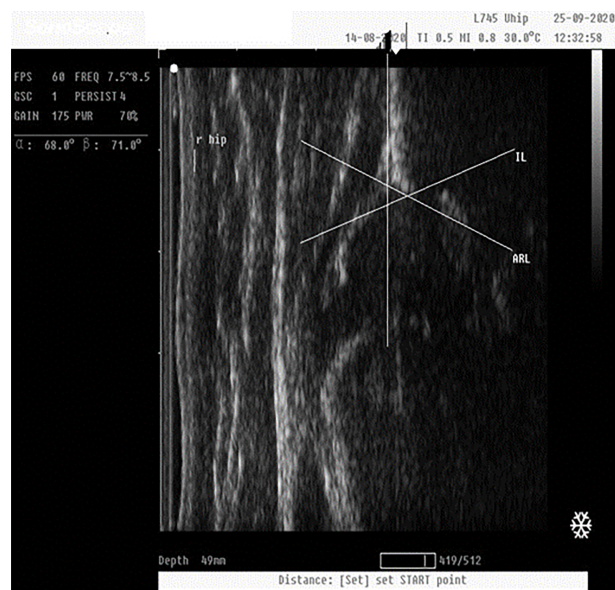
Type	Description	Bony roof	Bony rim	Cartilage roof	$\alpha$ -angle	$\beta$ -angle	Subtype
I	Mature hip	Good	Angular	Covers the femoral head	$\geq 60^\circ$	$< 77^\circ$	Ia: $\beta \leq 55^\circ$ Ib: $\beta > 55^\circ$
IIa	Physiologically immature hip (< 3 months)	Deficient	Blunt/Rounded	Covers the femoral head	$50^\circ$ to $59^\circ$	$> 55^\circ$	IIa <sup>+</sup> : $\alpha = 55^\circ$ to $59^\circ$ IIa <sup>-</sup> : $\alpha = 50^\circ$ to $54^\circ$ (at 6 weeks of age)
IIb	Delay of ossification (> 3 months)	Deficient	Rounded	Covers the femoral head	$50^\circ$ to $59^\circ$	$< 55^\circ$	—
IIc	Critical hip	Severely deficient	Rounded to flattened	Covers the femoral head	$43^\circ$ to $59^\circ$	$< 77^\circ$	IIc stable: $\beta < 77^\circ$ IIc unstable: $\beta > 77^\circ$ (under pressure)
D	Decentring hip	Severely deficient	Rounded to flattened	Displaced	$43^\circ$ to $59^\circ$	$> 77^\circ$	—
III	Decentring hip	Poor	Flattened	Pressed upward, perichondrium slopes cranially	$< 43^\circ$	$> 77^\circ$	IIIa: hypoechoic cartilage acetabular roof IIIb: hyperechoic cartilage acetabular roof
IV	Dislocated hip	Poor	Flattened	Pressed downward, perichondrium dips caudally	$< 43^\circ$	—	—

roof, flattened bony rim and pressed upwards cartilage roof by the dislocated femoral head. Type IV describes again femoral head dislocation. The bony roof is poor, the bony rim is flattened and the cartilage roof is pressed downwards from the dislocated femoral head. There is one exception, a type II joint with secondary ossification, with deficient bony roof, angular bony rim and cartilage roof that covers the femoral head. In this case, the hip is immature and second evaluation a few weeks later is needed.<sup>12,136</sup>

The above types may describe well the hip joint, but a more accurate classification system is needed in order to make a standardized treatment protocol and an easy communication system between different examiners. For these reasons, Prof. Graf developed the official classification for DDH, that is based

also on angles  $\alpha$  and  $\beta$ .<sup>138,139</sup> In this classification system, there are also types I to IV. Type I includes cases with the following angle measurements: angle  $\alpha$   $60^\circ$  or more. If angle  $\beta$  is  $< 55^\circ$ , the subtype is Ia, and if angle  $\beta$  is  $55^\circ$  or more, the subtype is Ib. Both subtypes are physiological (Fig. 4). Type II includes cases with angle  $\alpha$  between  $43^\circ$  and  $59^\circ$  and more specifically there are subtypes IIa (IIa<sup>+</sup> and IIa<sup>-</sup>), IIb, IIc, and D. Type IIa describes angle  $\alpha$  between  $50^\circ$  and  $59^\circ$ . In type IIa<sup>+</sup>, there is immature joint that seems to develop in the first 3 months and is a physiological condition, but in type IIa<sup>-</sup>, there is no such development in the first 3 months, and the hip should be treated (Fig. 5). In type IIb, the angle  $\alpha$  is between  $50^\circ$  and  $59^\circ$  in child older than 3 months, and the joint is defined as dysplastic and also needs treatment. Finally, type IIc (angle  $\alpha$  between  $43^\circ$





**FIG. 4:** Sonographic image of 43-day-old infant: alpha ( $\alpha$ ) angle 68°, type I in Graf classification, normal hip joint



**FIG. 5:** Sonographic image of a 2-day-old infant: alpha ( $\alpha$ ) angle 59° and beta ( $\beta$ ) angle 76°, type IIa in Graf classification, physiologically immature hip joint

and 49°) describes dysplastic acetabulum and nearly dislocated femoral head and treatment is needed. In type IIc cases, angle  $\beta$  is also important. If angle  $\alpha$  is

between 43° and 49° and angle  $\beta$  is more than 77°, then the joint is characterized as type D and it is the first stage of hip dislocation.<sup>136,140</sup>

Types III and IV include cases with angle  $\alpha$  less than 43° and describe infants with severe dislocation that requires also immediate action. In type III, the femoral head pushes upwards the cartilaginous roof. In type IV, the femoral head is completely dislocated posteriorly and upwards and pushes the cartilaginous roof downward.<sup>136</sup>

## VII. TREATMENT

Although DDH is a common musculoskeletal disease of young age, it is only in the last century that treatment techniques have been properly developed.<sup>14</sup> It is proved that the earlier the treatment begins, the better the results are. It is widely believed that after the age of 8–10 years old, the outcomes of any treatment technique are poor.<sup>141</sup>

The goal of treatment to restore the anatomy of the joint, by achieving a concentric reduction of the femoral head in the acetabulum. In order to gain good outcomes, it is needed to avoid complications as avascular necrosis and to correct the acetabular dysplasia, by having a proper stable and concentric position of the femoral head in the acetabulum.<sup>142–144</sup> There are conservative and surgical treatment techniques for DDH. Generally, when diagnosis is early and treatment starts immediately, conservative treatment is chosen. Surgical treatment with femoral or pelvic osteotomies is preferred when conservative treatment has failed (residual dysplasia) or in heavily dysplastic joints or neglected cases. During the follow-up, the patient has to be checked with ultrasound or radiographs regularly.<sup>131,145</sup>

Until the 1950s, DDH was usually diagnosed when children started to walk. It was Lorenz in late 1900s who first proposed the possibility of closed reduction and plastering in fixed maximal abduction and in 20th century, many other techniques have been developed.<sup>13</sup> In the 1950s, Pavlik proposed functional treatment for DDH, because of many cases of avascular necrosis of femoral hip that had been recorded with previous conservative treatment techniques.<sup>146–149</sup> Pavlik harness is a functional brace, used in infants with DDH and is now

considered as the elected conservative treatment (Fig. 6).<sup>150</sup> Historically, other similar devices have been used, such as the Frejka pillow and the LeDaman, Ortolani, and Lorenz devices, but literature has proven that Pavlik harness dominates with better results and lower incidence of complications.<sup>151</sup> The evolution in surgical conditions as antibiotics, blood transfusion and radiographs at the beginning of 20th century made open reductions possible and several femoral osteotomies (Salter, Pemberton, Chiari, and others) have been used.<sup>152</sup> Today, surgical treatment is rarely necessary.<sup>14</sup>

In developed countries, where ultrasonography of neonatal hip is used as the screening method, the majority of infants with DDH are treated immediately and conservative treatment is chosen, even in dysplastic cases.<sup>153</sup> The most useful and effective device is the Pavlik harness.<sup>147</sup> Pavlik harness is made by two shoulder straps that cross in the back and fast in a frontal thoracic belt. The legs of the infant are also held by two straps and the hips are flexed in more than 90° and abducted. This positions resembles the hip flexion of babies in utero and pushes the proximal femoral metaphysis with direction to the triradiate cartilage, leading to a concentric femoral head. The advantages of this device are that its



**FIG. 6:** A 6-week-old baby with a type IIc right hip joint treated in Pavlik harness

usage is atraumatic, it rarely causes avascular necrosis of the femoral head and it enables the patient to use its muscles.<sup>146,147,151</sup> Specifically, quadriceps, gluteal muscles, hamstrings, and abductors can have limited movement and play significant role in reducing hip dislocation.<sup>150,154</sup> Pavlik harness can be used more predictably in types II and III of Graf classification.<sup>155,156</sup> The duration of Pavlik harness usage is suggested to last at least 6 weeks and possibly for a complementary period of 6 more weeks, if there is development, but not total cure of acetabular dysplasia and non-concentric hip. There are studies that propose 3 months of usage when baby is younger than 3 months at the beginning of the treatment and for even longer if the baby is older than 4 months.<sup>157–159</sup> Ramsey et al. proposes duration of 9 months if the baby is between 3 and 9 months at the beginning.<sup>160</sup> It has also been suggested that prolonged period of Pavlik harness may cause avascular necrosis of the femoral head or delay the selection of alternative treatment processes.<sup>161</sup> Pavlik harness and almost every conservative treatment method need close follow-up with frequent ultrasonographic or radiological evaluation. The majority of authors believe that static, dynamic or combined ultrasonographic monitoring is reliable to evaluate if the joint construction develops with this certain device.<sup>114,162–168</sup> Pavlik harness is well studied and the outcomes are great, with success rates more than 50% to 99%.<sup>169–172</sup> Complications are rare, with avascular necrosis (rates from 0% to 28%) being the most common. Regarding infants with irreducible dislocations, no consensus has been reached among authors about the usability of Pavlik harness in infants with irreducible dislocations.<sup>83,142,173–176</sup>

Other similar devices, used when non-surgical treatment is chosen, are the Frejka pillow and the Rosen splint (mainly used in Nordic countries). In recent years, their usage is limited because of the high rates of complications. Skin irritation, pressure sores and avascular necrosis are the most common and in combination with Pavlik harness's better results they have almost been abandoned.<sup>151</sup>

For children older than 6 months, when DDH is diagnosed, in cases when Pavlik harness fails and for type IV (Graf classification) cases, closed reduction and spica cast remains the best treatment option.<sup>177</sup>



During the process, with the patient under general anesthesia, the physician reduces the non-concentric hip or both hips and stabilize them in flexed and abducted position. Then the spica cast is put and there is intraoperative radiological evaluation that certifies the successful reduction. In cases of unilateral disease, only the pathological limb is put in the spica and in cases of bilateral disease, both limbs are put in it (Fig. 7).<sup>178</sup> It has been proven that even a single-leg spica can provide adequate stability.<sup>179</sup> Hip spica has excellent results, but occasionally secondary operation is needed. The mean time of usage is 3–4 months and close follow-up with radiographs or ultrasound evaluation.<sup>177,180</sup> It is also needed to change the cast, because during the cast, the patient grows and the spica may harm soft tissues.

After the evaluation of ultrasonographic screening, the neglected cases have decreased and surgical treatment is mainly used in cases of conservative treatment failure. Femoral osteotomies are used to correct the anteversion and the valgus deformity of the femoral neck. For DDH, pelvic osteotomies, as Salter and Pemberton, together with open reduction of the femoral head are the elected procedures.<sup>181</sup> Salter osteotomy is an open wedge osteotomy, in which acetabular fragment is mobilized

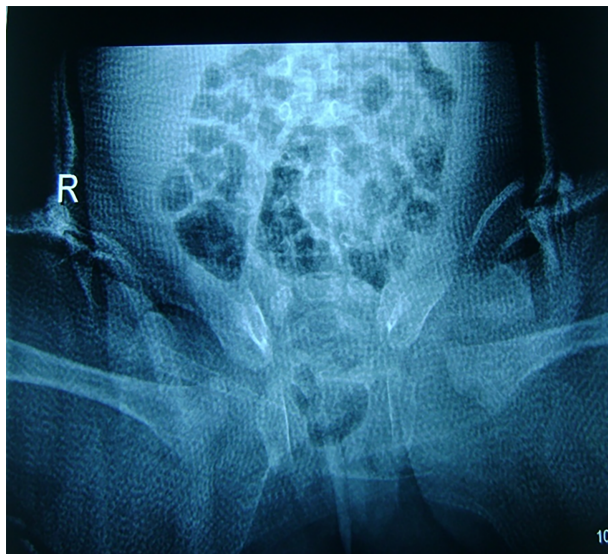
in order to cover the anterior side of femoral head. Symphysiolysis is a crucial stage of the procedure. Pemberton osteotomy is a transiliac osteotomy that re-shapes the joint.<sup>182–186</sup> These procedures are effective for children aged below 7 years old and may lead to severe complications as sciatic nerve damage, avascular necrosis of the femoral head, leg-length discrepancy, damage to the epiphyseal center and others. For children over 7 years old, there are other surgical procedures (open and closed triradiate cartilage) such as Dega, Steel, Tonis, and spherical osteotomies.<sup>187–190</sup> Finally, when all of these methods fail and concentric reduction cannot be achieved, there are salvage procedures such as Chiari medial displacement osteotomy and the Staheli procedure.<sup>191–192</sup>

## VIII. OUTCOME

DDH is the most common musculoskeletal disease of the infant age, known from the ancient years and causes severe degenerative lesions and clinical symptoms in adulthood, if it is not treated early.<sup>193</sup> The mean incidence in general population is 11.5/1000 live births with significant differences among different human races and it is recorded that girls are affected more often than boys and although the origin of this entity is not completely known, there are several risk factors as breech position in utero, family history, firstborn children swaddling in infant age and others that predispose for DDH presentation. The disease can be either unilateral or bilateral.<sup>16</sup>

The pathophysiology of DDH affects on bones and soft tissues causing dysplasia to dislocation of the hip joint. Specifically, the acetabulum grows abnormally and remains shallow and the femoral head grows in a non-anatomical position. The labrum cartilaginous roof and the ligaments are hypertrophied and the articular capsule is thick and stiff. The destruction and the slow growth of the femoral head epiphysis, because of the disease, leads to anatomic abnormality and aseptic necrosis.<sup>79</sup>

Physical examination is very important part of the diagnostic process for DDH. Every newborn has to be examined by a specialized physician for possible musculoskeletal problems in every part of the



**FIG. 7:** A 3-month-old baby with a type IV left hip joint reduced and held in hip spica

body. Barlow and Ortolani maneuvers are appropriate and in case of positive sign, there are strong indications for presence of DDH.<sup>100,101</sup> When DDH is misdiagnosed, children in walking age may develop Trendelenburg gait and for adults with undiagnosed disease signs and symptoms of OA appear in young age.<sup>194</sup> Although physical examination gives information for the possibility of DDH, nowadays the gold standard diagnostic approach is the usage of ultrasonography to evaluate the infant hip and in many western societies is part of the basic screening test of the newborns.<sup>195</sup> The most popular and useful technique is the Graf method, developed by Prof. Reinhardt Graf in early 1980s.<sup>12</sup> This method enables the physician to examine the hip joint of the infant based on the anatomical structures by measuring two angles ( $\alpha$  and  $\beta$ ) that show the coverage of femoral head by osseous and labral acetabulum. According to this angles, the infant hips are classified with Graf classification in types I to IV, from normal joint, to immature joint, subluxation and dislocation. The classification is vital because each different type leads to different and specific treatment approach. In the literature, 4–6 weeks old is defined as the best age for the infant to be examined with ultrasonography.<sup>89</sup>

Radiographs are useful for children with DDH at the age of 4–6 months or older and facts as Acetabular Index and Shenton line are pathological.<sup>129</sup> Radiography is also used during the follow-up of children treated with Pavlik harness or hip spica.<sup>131</sup> CT scan and MRI are not widely used for the evaluation and diagnosis of this entity.<sup>133–135</sup>

The treatment of DDH is connected with the necessity of early diagnosis. After the wide usage of ultrasonography less operative means of treatment have dominated. The goal of treatment is to restore the anatomy of the joint, by achieving a concentric reduction of the femoral head in the acetabulum.<sup>142</sup> The treatment is important to begin immediately after diagnosis is confirmed. Closed reduction and devices as Pavlik harness or hip spica are used for children less than 3 months with excellent outcomes. For children older than 6 months and until the walking age, hip spica remains the best option for treatment. Nowadays, with the widespread usage of ultrasonography, the neglected or misdiagnosed

cases have decreased and almost disappeared in developed countries. For these cases, operative techniques as femoral and pelvic osteotomies are available, but the results are not always satisfactory. Generally, after the age of 8–10 years, the results of every treatment option are very poor.<sup>141,196</sup>

The frequent incidence of DDH should make the physicians to be aware and careful during the clinical and sonographic evaluation, because misdiagnosed disease will cause severe consequences to the patient.<sup>197</sup> The extensive usage of hip ultrasonography is important and should be used as screening test in every infant being born, because in this way the children with DDH can be easily diagnosed and get treated. Management requires the proper option to be chosen according to the types I to IV (Graf classification) and to evaluate the child closely with close clinical and radiological monitoring.

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