Variations in developmental dysplasia of the hip – current concepts

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Abstract
Developmental dysplasia of the hip (DDH) is one of the most common congenital defects with an incidence of around 2 cases per 1000 births, affecting the development of the acetabulum and the femoral head. Etiopathogenesis remains unclear but certain risk factors have been associated with DDH including the female gender, postmaturity, primiparity, oligohydramnios and breech presentation. If left untreated, hip dysplasia is considered a main cause of early osteoarthritis. Reviewing the literature, the purpose of the current study is to present current evidence regarding the anatomical abnormalities of DDH during infancy, childhood and adulthood. Using the searching tools on the internet, a thorough search, evaluation and selection of recent published articles in reliable international electronic libraries was conducted regarding the anatomical variations in developmental dysplasia of the hip. The results, extracted from these manuscripts, were the basic source of the current study. It was found that hip anatomy is affected in many different ways which range from sole acetabular dysplasia and stability to subluxation and dislocation. Acetabular defects along with femoral head and soft tissue abnormalities are usually present, making surgery a very challenging process. Dominant symptoms in infancy involve length discrepancy and limited abduction while groin pain and abnormal gait appear later in life. In cases of early diagnosis, DDH can be treated efficiently. Therefore, careful examination of all infants should be performed using the Barlow and Ortolani tests along with ultrasound, when there is a high clinical suspicion. It would be advisable though to screen all infants in order to avoid delayed diagnosis. In later life, radiographs are considered the primary diagnostic tool, whereas a considerable advancement to treating adult DDH is the growing use of personalized (custom-made) implants.

Keywords: Developmental dysplasia; Hip; Anatomical variations; Dislocation

1. Introduction
Developmental dysplasia of the hip (DDH) is a common congenital defect which might affect the development of both the femoral head and the acetabulum [1, 2]. DDH was formerly known as congenital dislocation of the hip (CDH) but nowadays this term has been abandoned since abnormal hip anatomy can be present without a dislocation [3]. It is characterized by multiple clinical and anatomical manifestations rendering it a very complex disorder, which might be asymptomatic in early stages [1, 4]. Later in life, if left untreated, DDH is considered to be the main cause of early osteoarthritis due to alterations of normal biomechanical stresses that could result in physical disability and functional disorder [5, 6]. In particular, total hip arthroplasty (THA) in adults younger than 60 years old, is frequently attributed to pre-existing DDH [7].

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Regarding epidemiological data, its incidence is estimated at around 2 per 1000 births and there is also a tendency for affecting the female gender, 4 times more likely, as well as the left hip (60%) [8, 9]. However, since many countries have adopted selective ultrasound screening, a lower incidence of DDH has been recorded [8]. Concerning the etiopathogenesis, multiple factors have been associated with the development of the disease including breech presentation, oligohydramnios, postmaturity and primiparity [10, 11]. Genetic factors also play an important but still unidentified role as it has been found that several genes, involved in osteogenesis and chondrogenesis, have been related with the onset of DDH and in cases of familial DDH, first degree relatives are estimated to have more than 10-fold increase in the occurrence of DDH [12, 13]. In addition, a strong correlation has been observed between DDH and certain other abnormalities including metatarsus adductus, congenital muscular torticollis, congenital knee dislocation and Down syndrome, thus the term syndromic DDH [11, 14].

The purpose of the present review was to demonstrate current evidence regarding anatomical abnormalities of DDH during three developmental stages of life (infancy, childhood and adulthood).

2. Material and methods

A thorough review of the literature was conducted in order to present recent data regarding the anatomical variations of developmental dysplasia of the hip during infancy, childhood and adulthood. The material included studies that were original articles written in the English language, involving randomized controlled trials, prospective or retrospective comparative studies, systematic reviews, meta-analyses and case series. On the contrary published articles such as letters to the editor, editorials, comments, expert opinions, technical notes, case reports or book chapters were excluded from the study.

The terms “developmental dysplasia of the hip” or “congenital hip dislocation” in combination with “anatomical variations”, “abnormalities”, “infancy”, “childhood” and “adulthood” were used as keywords in order to search for relevant studies in popular library databases like Medline/Pubmed, Scopus, ScienceDirect and Cochrane Library.

The articles that met the inclusion criteria were reviewed closely so as to select for our study, the most recent manuscripts and some primary older articles. However, no quantitative or systematic analysis of data was conducted, due to the high level of heterogeneity and significant discrepancies among the included studies.

3. Results

3.1 Anatomical variations in DDH in infancy and childhood

In infancy, DDH may occur with a spectrum of anatomical variations ranging from a stable hip with acetabular dysplasia to an unstable hip with subluxation or dislocation of the femoral head [9]. In rare cases the dislocation may be irreducible on neonatal examination predisposing to a teratologic dislocation that appears in intrauterine life, is related to neuromuscular conditions or genetic disorders and leads to a resistant to closed reduction and stiff hip, during infancy [15]. In such cases a false acetabulum (pseudoacetabulum) might be already present at a very early stage which is a sign of established prenatal hip dysplasia [16].

Two distinctive lesions named “limbus” and “neolimbus” have been reported in patients with DDH due to superior migration of the femoral head causing abnormal contact stresses [17, 18]. The term limbus refers to a thickened labrum that is hypertrophied with fibrous and fibrocartilaginous overgrowth and has the potential to become inverted and mechanically block the concentric reduction of the dysplastic hip [17]. The neolimbus is defined as a hypertrophied ridge of cartilage in the superolateral region of the acetabulum, which arises beneath the limbus, because of the constant strain from the dislocated hip [18].

Apart from limbus and neolimbus, a variety of other anatomical variations has been observed in infancy and early childhood, that embraces capsule constriction, inverted labrum, hypertrophic ligamentum teres, contracted transverse acetabular ligament, excessive fibrofatty tissue filling the acetabulum (pulvinar) or flattening of the femoral head [17, 19]. When these structures obstruct reduction, they have to be excised surgically [20].

An early diagnosis of DDH is very important as at very early stages, there is a high chance of treatment without surgery [21, 22]. Diligent examination should take place during infancy and early childhood in order to avoid late neglected cases [22]. Limited hip abduction and lower limb length discrepancies are two major clinical signs that should alert doctors [9]. In general, screening should start at birth when the Ortolani and Barlow tests should be performed [6]. Both
of them have high specificity but low sensitivity, thus some cases of true DDH can be missed especially when there is only mild dysplasia of the acetabulum or the hip is permanently dislocated [9, 23].

Therefore, in cases of high clinical suspicion, like after positive physical examination, ultrasound (US) should be used to diagnose DDH, which represents the primary imaging method from birth to the age of 4-5 months [24]. In particular, two angles are measured and the patient is classified into 4 groups according to Graf [25]. The alpha angle is formed by lines along the ilium and the bony acetabulum and represents the depth of the acetabulum whereas the beta angle is created by lines along the ilium and the labrum [4, 21, 25]. Values of alpha angle, more than 60 degrees and beta angle, less than 55 degrees, are considered as normal (Table 1).

In infants older than 4-6 months and young children, radiographs are indicated to detect DDH [9]. At this stage, the femoral head begins to ossify which makes radiographic examination suitable [26]. Certain measurements including the Hilgenreiner’s line, the Perkin’s line, the Shenton’s line, the acetabular index (AI) and the center-edge angle (CEA) of Wiberg, are taken into consideration in the anteroposterior (AP) view of pelvis [4, 6].

The Hilgenreiner’s line is horizontal line through the right and left triradiate cartilage and the femoral head ossification should be inferior to this line. Otherwise there is a high possibility for DDH [22]. The Perkin’s line is perpendicular to Hilgenreiner’s line through a point at the lateral margin of the acetabulum and in cases where the femoral head ossification is not medial to this line, DDH should be further investigated [22]. The Shenton’s line represents an arc along the inferior border of the femoral neck and the superior margin of the obturator foramen which should be always intact and continuous [22]. The AI is defined as the angle between the Hilgenreiner’s line and a line tangentially connecting the inferior margin of the iliac bone and the superolateral part of the acetabular bony rim, assessing the inclination of the acetabulum that should be less than 25° in patients older than 6 months [22]. Finally, the CEA of Wiberg is an angle between Perkin’s line and a line from the center of the femoral head to the lateral edge of the acetabulum. It is considered reliable only in children over 5 years old and depicts the lateral coverage of the femoral head. Values greater than 20° are referred as normal [27].

Table 1 Ultrasound classification system for DDH in infants according to Graf

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
<th>Bony roof</th>
<th>Bony rim</th>
<th>Cartilage roof</th>
<th>Alpha angle (degrees)</th>
<th>Beta angle (degrees)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Mature hip</td>
<td>Good</td>
<td>Angular/blunt</td>
<td>Covers the femoral head</td>
<td>≥ 60°</td>
<td>&lt;77° la&lt;55° lb&gt;55°</td>
</tr>
<tr>
<td>IIa</td>
<td>Physiological (&lt; 3 months)</td>
<td>Deficient</td>
<td>Rounded</td>
<td>Covers the femoral head</td>
<td>50-59°</td>
<td>&gt;55°</td>
</tr>
<tr>
<td>IIb</td>
<td>Delay of ossification (&gt;3 months)</td>
<td>Deficient</td>
<td>Rounded</td>
<td>Covers the femoral head</td>
<td>50-59°</td>
<td>&lt;55°</td>
</tr>
<tr>
<td>IIc</td>
<td>Critical hip</td>
<td>Severely deficient</td>
<td>Rounded to flattened</td>
<td>Still covers the femoral head</td>
<td>43-49°</td>
<td>&lt;77°</td>
</tr>
<tr>
<td>IId</td>
<td>Decentering hip</td>
<td>Severely deficient</td>
<td>Rounded to flattened</td>
<td>Displaced</td>
<td>43-49°</td>
<td>&gt;77°</td>
</tr>
<tr>
<td>III</td>
<td>Dislocated hip</td>
<td>Poor</td>
<td>Flattened</td>
<td>Pressed upward, perichondrium slopes cranially</td>
<td>&lt;43°</td>
<td>&gt;77°</td>
</tr>
<tr>
<td>IV</td>
<td>Dislocated hip</td>
<td>Poor</td>
<td>Flattened</td>
<td>Pressed downward, perichondrium is horizontal or dips caudally</td>
<td>&lt;43°</td>
<td></td>
</tr>
</tbody>
</table>
The treatment should start as soon as the diagnosis of DDH is set. In general, the risk, complexity and upcoming hip complications are impaired with delays in diagnosis and early treatment [6, 29]. In infants aged less than 6 months, abduction splinting with Pavlik harness could be applied when the hip is reducible. There is a strong contraindication for such a bracing in patients with a teratologic hip dislocation or severe spasticity [6, 28, 29]. With the Pavlik harness the hip is positioning in 90°-100° of flexion with the anterior straps, whilst the posterior straps prevent adduction of the hips facilitating early concentric reduction in order to prevent degenerations in the joint. The reduction should be checked with ultrasound or radiograph every 4-6 weeks [6, 28, 29].

This type of cast in order to be effective (80-100%), requires a 23 hours/day constant use for at least 6 weeks and afterwards application only during the night for further 6-8 weeks until findings of normal hip anatomy can be observed [29]. Caution should be taken in order to avoid avascular necrosis (AVN) due to impingement of the posterosuperior retinacular branch of the medial femoral circumflex artery which can occur when hip abduction exceeds 60° (1-8%) [30]. Other severe complications that have been reported after Pavlik harness usage, involve potential transient femoral nerve palsy due to extreme hip flexion, or erosion of the pelvis superior to the acetabulum and abnormal development of the posterior wall [6, 28].

When the initial diagnosis of DDH is performed in ages between 6 and 18 months or in cases where Pavlik treatment fails, closed reduction is proposed, under general anesthesia, followed by use of spica casting for 3 months. Commonly, the hip is immobilized in 100° of flexion, 45° of abduction and neutral rotation, whereas the cast should be changed every 6 weeks [6]. However for patients with unsuccessful closed reduction or age more than 18 months, open reduction and postoperative management with spica cast is typically proposed [6]. Furthermore, in patients with residual hip dysplasia, age over 2 years old and anatomic changes on the femur, like coxa valga, open reduction should be combined with femoral osteotomy (frequently until 4 years old) [16, 31]. On the contrary, in children, aged over 4 years old, with severe hip dysplasia and anatomic changes on the acetabulum, open reduction are preferably followed by pelvic osteotomies [31, 32].

### 3.2 Anatomical variations in DDH in adulthood

Anatomical abnormalities of DDH in this stage of life may vary as well. However, certain common characteristics have been reported [9]. In particular, in the majority of cases, the acetabulum is shallow, lacking in anterolateral and superior coverage [33]. Additionally, a false acetabulum might be formed if the hip is dislocated. Regarding the femoral side, disorders such as increased femoral anteversion, coxa valga, shorter and smaller femoral neck, posterior displacement of the greater trochanter, aspherical femoral head and hypoplasia of the femoral intramedullary canal may also be observed [33, 34].

These femoral malformations have been directly related with femoroacetabular impingement which is common in DDH and aggravates symptoms [35]. Moreover, in radiographs, the femur can be demonstrated with narrower and straighter intramedullary canal, thinner cortical diameter and abnormal anterior bowing [34, 36]. Soft tissue abnormalities can be also apparent and might concern capsular contracture, labral thickening along with muscular shortening and insufficiency due to the altered position of the femoral head and mechanically different contact stresses [37].

Although various classifications have been referred for DDH in adulthood, those reported by Crowe and Hartofilakidis have prevailed, especially when surgery is considered. Crowe classified DDH into 4 grades according to femoral head subluxation, whereas Hartofilakidis identified 3 types taking into consideration the acetabular deformity [38, 39] (Tables 2 and 3).

**Table 2 Crowe classification for DDH in adults**

<table>
<thead>
<tr>
<th>Crowe Classification</th>
<th>Description</th>
</tr>
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<tbody>
<tr>
<td>I</td>
<td>&lt;50% subluxation or proximal displacement &lt;10% of the pelvic height</td>
</tr>
<tr>
<td>II</td>
<td>50-75% subluxation or proximal displacement 10-15% of the pelvic height</td>
</tr>
<tr>
<td>III</td>
<td>75-100% subluxation or proximal displacement 15-20% of the pelvic height</td>
</tr>
<tr>
<td>IV</td>
<td>&gt;100% subluxation or proximal displacement &gt;20% of the pelvic height</td>
</tr>
</tbody>
</table>
Table 3 Hartofilakidis classification for DDH in adults

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dysplasia (Type A)</td>
<td>- Femoral head within the acetabulum but subluxated</td>
</tr>
<tr>
<td></td>
<td>- Superior segmental defect of the acetabulum</td>
</tr>
<tr>
<td>Low Dislocation (Type B)</td>
<td>- Femoral head within a false acetabulum superiorly which partially covers the true acetabulum</td>
</tr>
<tr>
<td></td>
<td>- Absence of the superior wall</td>
</tr>
<tr>
<td>High Dislocation (Type C)</td>
<td>- Superior and posterior migration of the femoral head</td>
</tr>
<tr>
<td></td>
<td>- No contact with true acetabulum</td>
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</tbody>
</table>

The main symptomatology in this stage of life includes groin pain exacerbated by physical activity and Trendelenburg gait, which is attributed to abductor insufficiency or fatigue and established arthritic lesions [33]. Neglected cases with hip dislocation which are left untreated may have more obvious symptoms like limping or length discrepancy [4]. The diagnosis of DDH is confirmed by radiographs (AP, lateral and false-profile view) but a CT (Computed Tomography) scan may be useful preoperatively for a detailed analysis of hip anatomy [7]. Certain parameters are taken into account including the Tönnis angle, the lateral center-edge angle (LCEA) of Wiberg and the anterior center-edge angle (ACEA) of Lequesne [40].

The Tönnis angle is formed between a horizontal line and a line along the superior acetabulum, depicting the acetabular inclination on the AP view. Values less than 10° are considered as normal [40]. The LCEA is created between a vertical line through the center of the femoral head and a line that connects the lateral edge of the acetabulum with the femoral head center, assessing the superolateral coverage of the femoral head on the AP view [41]. Values between 25° and 39° are considered within the normal range. The ACEA is shaped between a vertical line through the center of the femoral head and a line which joins that anterior edge of the acetabulum with the femoral head center, assessing the anterior coverage of the femoral head on the false-profile view [7]. Values of ACEA within 25-40° are regarded as normal.

The role of long-term nonsurgical treatment in symptomatic DDH is limited and generally ineffective leading to progression of secondary hip osteoarthritis [1, 33]. Operatively, hip arthroscopy which has been developed over the last years might enhance the quality of life in patients with mild dysplasia and chondral, labral or proximal femoral cam-type lesions. However this surgical technique is contraindicated in moderate to severe dysplasia as at these stages the underlying pathology cannot be corrected due to severe osseous instability [1, 20, 33]. Periarticular or salvage osteotomies have also been used in symptomatic hip dysplasia of young adults displaying in general, limited indications (contraindication to severe DDH), high morbidity and moderate outcomes [1, 33].

In patients with end-stage osteoarthritis secondary to DDH, the treatment of choice is the total hip arthroplasty (THA) that relieves from pain and improves the quality of life. Technically the surgeon aims to place the acetabular component in the true acetabulum to restore normal hip kinematics. This may cause considerable leg lengthening, which subsequently might require femoral shortening. Care should be taken to avoid sciatic nerve palsy since in cases where lengthening exceeds 4 cm, there is an increased risk of neurovascular injury. Furthermore, it should be mentioned that the revision rate for THA after DDH is higher as compared to non-dysplastic hips [7, 12, 37].

4. Discussion

Hip dysplasia directly affects hip function. In infancy, a dislocated hip, with severe dysplasia that would be left untreated, will disturb the normal anatomical growth due to abnormal mechanical stresses, evolving into a shallow acetabulum and abnormal femoral head [7, 33]. This would have a direct impact on hip’s range of motion and length of the lower extremity leading to Trendelenburg gait or limping [22]. On the contrary, a stable and non-displaced hip with mild dysplasia might be asymptomatic for many years, as commonly, the intraarticular damage demonstrates a slow and gradual aggravation due to hip’s relative stability and thus symptomatology presents later in life [4, 9, 33].

In the past years, the hip dysplasia that was diagnosed in adolescence and adulthood was attributed to a delayed diagnosis of DDH which had theoretically appeared in infancy [42]. Nowadays, it is believed that since the acetabulum continues to develop until adolescence, it could not be excluded that dysplasia in adolescents may be initially diagnosed,
without formerly existing [33, 43]. Thus, adolescent-onset acetabular dysplasia might be a separate disease with similar symptomatology and clinical findings [42, 44]. In addition, there are cases in the literature of residual hip dysplasia presenting later in life [45, 46]. Modaresi et al. concluded that, even after prompt diagnosis and early treatment in infancy and early childhood, DDH may relapse [47]. Therefore, systematic monitoring of these patients should take place in order to avoid early symptomatology and arthritis [46].

As mentioned above, the diagnosis of DDH is made at infancy and early childhood with the use of physical examination and US or Xrays [9]. However, there is a controversy regarding a universal or selective US screening program [9]. Several countries have adopted a universal screening increasing the overall cost, but it does not seem to prevail over the selective assessment [48]. This is probably because of the variability of all US metrics even of the alpha angle, which still remains the least variable and most widely used metric, making “true” DDH incidence uncertain [24]. An ongoing debate about the time for US screening exists as well. The ideal time has yet to be established reaching even 6 months in some studies, despite the study of Gokhaman et al. that US performed at 8 weeks can provide safe and reliable results [21].

While radiographs, ultrasound and CT scan are typically used to diagnose DDH, magnetic resonance imaging (MRI) might rarely be used to determine concentric reduction of the hip after manipulation, or to delineate the soft issues that block reduction [2]. One distinctive feature of dysplastic hips that MRI can reveal is the lower cartilage surface [38]. This factor along with the altered joint mechanical stresses and different structure of articular cartilage in DDH that Feng et al. have reported, might be the primary reasons for early arthritis [49].

Due to the complexity of the disease, several authors have provided classifications for DDH, including Crowe, Hartofilakidis, Gaston, Eftekhar and Kerboul [38, 39, 50-53]. Eftekhar’s and Kerboul’s classifications are based on the grade of dislocation of the femoral head and the anteroposterior position of the femoral head, respectively [50]. Whereas they have displayed good interobserver and intraobserver reliability, they are not as competent as Crowe’s and Hartofilakidis classifications in popularity, which are considered as the most reliable and thus they are commonly used [50-52]. However, these two popular classifications have certain disadvantages. Crowe’s grades do not take into consideration dysplastic hip anatomy while Hartofilakidis types might not be so reliable between less experienced doctors [38, 39]. On the other hand Gaston et al. provided another classification which is advocated as predictable for technical problems during total hip arthroplasty [53].

Apart from hip anatomical degenerations, DDH has been associated with concomitant alterations on knee anatomy due to modified biomechanics [54, 55]. Disorders such as valgus knee, smaller femoral condyles, shallower trochlear groove and patellar instability, have been found in patients with DDH [54]. Particularly, valgus on the alignment in lower extremity has been positively correlated with lateral hip migration, which should be assessed prior to hip surgery [55]. Patellar malalignment might also predispose to knee pain and arthritis which can be misleading and draw doctor’s attention away from the hip joint [39]. In parallel, hip pathology might be related to spine pathology, as Segreto et al depicted a positive correlation between early onset or adolescent idiopathic scoliosis and DDH [56]. Nevertheless, more studies are required in this field in order to have more reliable results.

5. Conclusion

DDH is a complex disorder with unclear pathogenesis which can become symptomatic at almost any stage of life. Dysplasia mainly starts from infancy to adolescence when there is a potential for normal growth and development, but sporadically this dysfunction might become apparent even in adulthood. Dysplastic hip anatomy is unique for every patient with a diverse range of anatomical abnormalities. However, certain recurrent trends have been observed.

Moreover, it should be elucidated that most people with untreated DDH will end up with hip osteoarthritis which makes prompt diagnosis imperative. Further studies focusing on the manifold anatomy of the dysplastic hip should be conducted in order to facilitate diagnosis, better understand the pathogenesis and individualize treatment options.

Compliance with ethical standards

Disclosure of conflict of interest

All authors declare that they have no competing interests.
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