Chapter 1

Introductory Chapter: Five-Dimensional Approach to the Developmental Dysplasia of the Hip

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Additional information is available at the end of the chapter

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Developmental dysplasia of the hip (DDH) is not a specific disorder; it is rather a scale of overlapping and transforming conditions. It ranges from occult dysplasia seen on ultrasound screening of newborns, neonatal hip instability and dislocated hip whether reducible by orthopaedic manipulation or not. The hallmark of DDH is acetabular dysplasia—abnormality in size, shape or orientation of acetabulum. A thoughtful elucidation regarding DDH is that it is 'a common and preventable cause of childhood disability' [1]. Complications and consequences of DDH make this time interval much longer, though.

The incidence of many faces of DDH is fortunately distributed: it is reported to be as much as 8% for dysplasia, 1–3% for neonatal hip instability and <0.2% for frank dislocation [2–4]. These epidemiological data are greatly influenced by both diagnostic criteria and diagnostic methods used [5, 6]. They evolve not only due to demographic changes of population, predominantly through migrations and genetic mixing, but also due to changes in nutrition [7–9].

DDH is not a disease of modern age. It was recognised and described by Hippocrates as a congenital dislocation of the hip. Dysplastic hips and presence of false acetabulum were found in skeletons from medieval times [10–14].

Present etiological concept of DDH is multifactorial, consisting of endogenous (genetic disorders of collagen or collagen-related enzymes, transmembrane G-protein) [15, 16] and exogenous factors (related to intrauterine biomechanics, such as breech position and history of prior pregnancies, or environmental like birth in a certain season, swaddling technique) [17–19]. Breech presentation, positive family history of DDH, female sex, vaginal delivery, primiparity and oligohydramnion are usually regarded to as DDH risk factors. Some authors include other mechanical intrauterine restrictions (large baby, multiple pregnancy), advanced maternal age and delivery-related conditions (post-maturity) [1, 20]. It is worth noting that premature birth is not a risk factor for DDH [21]. Risk factors have limited clinical importance, however, due to both low sensitivity (10–27% of all infants who have DDH also have any risk factor) and low specificity (under 10% of children with risk factors have DDH) [22, 23].

Historically, there were several crucial events that improved both the diagnosing and treating this disease.

- First, understanding the hip biomechanics, Lorenz in 1895 introduced first successful method of closed reduction, using plaster cast in extreme abduction for retention [24]. Results were immediately supported with new method discovered same year: X-rays, and so the Lorenz method became widespread.
- Then, in 1935, Italian paediatrician Ortolani established a diagnostic manoeuvre used to verify present dislocation with audible and sensible 'click' [25]. He was the first to recognize the importance of diagnosis of dislocated hip in infants. A systematic screening of newborn hips has started.
- In 1944, Pavlik begun applying the harness as means for keeping dysplastic hips mobile but limited to advantageous abduction angles, thus promoting biomechanical stimulation of normal hip development [26].
- Modern operative treatment of acetabular dysplasia begun with Chiari [27] and Salter [28] pelvic osteotomy.
- In 1961, Charnley introduced modern concept of total hip replacement in the treatment of osteoarthritis, a common sequela of hip dysplasia in adult age [29].
- Following the technological improvements, Graf introduced ultrasound as a method for visualisation of the hip and described diagnostic criteria for assessment of hip dysplasia [30].
- Finally, with screening data available, Klisic introduced a new name 'developmental dysplasia' [31].

Basically, there are five very important dilemmas that demarcate the struggle with this rather recalcitrant medical condition. Their analysis reflects both the complexity of problem and diversity of solutions currently available across the medical practice in the whole world.

1. Screening for hip dysplasia: overlooking versus overtreating

Neonatal hip joint demonstrates significant potential for growth and remodelling. Still, the outcome of nonoperative reduction of dislocated femoral head and its safe containment within the acetabulum strongly depends on timing. If a treatment begins within first 7 weeks, it will be highly successful [32–35] regardless if one or both hips are treated [36]. That is why meticulous clinical examination of hips in newborns is mandatory for decades. Establishing a diagnosis of DDH after 3 months of age is considered as a late presentation, with estimated incidence from 0.02 to 0.2% [37, 38]. It is associated with higher rate of operative treatment, worse prognosis and increased healthcare cost [39–43].

Unstable or dislocated hip is usually diagnosed by combined Ortolani-Barlow manoeuvre, with satisfactory specificity (>84%) but controversial sensitivity (from 7 to 98% in various studies) [44]. These clinical signs, however, cannot pinpoint acetabular dysplasia. For that reason,

in some medical systems, an ultrasound screening is also mandatory and universal [8], while in others, it is used only in selected, targeted cases [45]. These variations in screening protocol are due to economic, organisational reasons, as well as the concern of over diagnosing and possible unnecessary treatment [46–48]. Most common ultrasound screening methods are according to Graf, Harcke, Terjesen and Suzuki. Data from Austria, UK and Ireland suggest that universal ultrasonic screening for DDH reduced both overall average healthcare expenses and the need for operative treatment [2, 45, 49–52], although there are different opinions in the USA [53].

While very valuable for early detection of hip dislocation, and without any absolute contraindications [54] ultrasound in first 2 weeks of life has limited sensitivity to detect clinically relevant dysplasia, since a fraction of newborns have underdeveloped but healthy hips—a temporarily false positive result [4, 55, 56]. Some authors even suggest that ultrasound in first 6 weeks should confirm the diagnosis of DDH only if hip is decentred (Graf III type) or dislocated (Graf IV type). For true incidence of hip dysplasia, a correlation of ultrasound data, clinical examination and the number of late presented cases requiring operative treatment should all be analysed.

Nevertheless, the problem how to discriminate between dysplastic hips and healthy hips still remains—ultrasound is too dependent on examiner's skills, while radiographic criteria are usually biased by pelvic rotation [57]. Effective screening for DDH should be characterised by low percentage of cases that require surgical intervention, and all of those due to failures of nonoperative treatment, rather than due to late detection [49, 58]. In some studies, the majority of patients with symptomatic dysplasia in adult age did not meet criteria for selective ultrasound screening in infant age—they were false negative on clinical examination [59].

2. Neonatal hip instability: nature versus therapy

Like hip dysplasia, neonatal hip instability follows similar diagnostic concerns. This condition is diagnosed either by provocative tests (Barlow) or by dynamic ultrasound testing (Harcke technique) [60] with substantial reproducibility and accuracy only achieved in combination of these methods [40]. On the other hand, failure of recognition and treatment of neonatal hip instability can lead to significant hip dysfunction [61].

Neonates are usually born with slight flexion contracture (25–30°), which should spontaneously decrease to <20° at 6 weeks, and 7° at 12 weeks. In addition, one should bear in mind that majority of hips clinically unstable at birth will resolve spontaneously within first 8 weeks [62], in some cases until 3 years of age [63]. In other words, specificity of clinical and ultrasonic examination improves with growth. This is particularly true for testing if there is limited abduction, which meets its peak of reliability as DDH marker at the age between 3 and 6 months [64].

The relation between abduction position and movements and proper stimulation of dysplastic/unstable acetabulum to become better is clearly demonstrated, and positioning of legs influences the outcome of hip development [65]. While wide (double) diapering stimulates beneficial

dynamic abduction of both hips, there is also an opposite praxis of swaddling (hips in extension and in zero abduction) either due to traditional routine in some parts of the world (Middle East, Japan, Native Americans, etc.) [66–68] or for the prevention of excessive crying and promoting sleep [69]. It is clear, however, that risk for hip deterioration if legs are kept laced grossly surpasses all potential benefits, which are easily achievable by other, less hazardous means.

3. Natural history of DDH: prevention versus operation

We already stated that dysplastic and unstable hips may undergo spontaneous recovery [70]. As for the cohort of non-recovering hips, it has been observed that DDH leads to significant loss of normal joint function [71]. Dysplastic hips have tendency to evolve over years into painful and debilitating osteoarthritis [72–74], while dislocated hips are accompanied with short posture and waddling gait throughout life, and if not reduced operatively within the first 8 years, painful syndrome may eventually develop [59, 75]. In patients with untreated unilateral dislocation, pelvic obliquity deteriorates the distribution of hip force on contralateral hip joint, contributing to degeneration on that side as well, along with further compensatory dysfunctions of trunk and knees [11].

DDH and osteoarthritis share genetic biomechanical etiological aspects [76]. Longitudinal studies have revealed that degenerative changes induced by hip dysplasia develop more rapidly than in other predisposing conditions [77]. Total hip replacement (THR) is a surgical procedure that is most often performed in treating symptomatic advanced osteoarthritis, especially in younger age [78–82]. The diagnosis of DDH in first-order relative increases a chance for THR by the age of 65 [83]. Recent studies show that average hospital cost for primary THR secondary to DDH is higher than in other cases. Also, the severity of DDH additionally increases those expenses [79, 84]. If DDH is diagnosed early and the treatment was nonoperative, the rate of osteoarthritis at long-term follow-up is twice lower than after open reduction [81]. On the other hand, survival rate of dislocated hips that undergone operative treatment in infancy including innominate osteotomy was 54% at the age of 45 [85].

4. The follow-up challenge: hip morphology versus function

Since DDH is a kind of 'moving target' throughout patient's life, several assessment protocols are in use for follow-up once the diagnosis is established, depending on the kind of intervention (observation, nonoperative or operative procedure), age and complaints. They all have the same two prominent characteristics:

(a) Low reliability and inter-observer concordance [86].

(b) Inadequate correlation of functional and radiographic results [87], implying that not all radiographically dysplastic/arthritic hip joints are the same, and that more than morphologic factors influence the onset and severity of symptoms.

Health-related part of elusive term we refer to as 'quality of life' (QoL) includes, but is not limited to, satisfaction in physical, emotional and social aspects of life. Quality of life with DDH is mostly affected by pain, gait disturbance, limited range of motion and leg length discrepancy. These factors are not independent, they aggravate each other. Patients become regular consumers of various healthcare services and products, spending days and money on rehabilitation, usually getting weight because of inadequate activity. Several studies demonstrated long-term improvement in QoL after THA in patients diagnosed with DDH [88, 89]. Although very important for patient, QoL assessment should be primarily used to identify their expectation regarding the type of treatment and should not replace clinical examination and standard diagnostic methods [90, 91].

5. The impact of DDH on healthcare: cause versus effect

Many diseases have been imposing a strong burden to healthcare service on global scale, in aspects of organisation, cost and consequences of diagnostic and therapeutic modalities indicated. In a rather long list, one could count in tuberculosis, diabetes, cardiac failure, cancer, AiDS and DDH. Most of them share the same characteristic of significant mortality, direct or indirect through complications. On the other hand, DDH is among the few exceptions that are not directly life threatening but deteriorate the quality of life and/or working ability up to great extent and for a long time [92].

Osteoarthritis is one of the major causes of non-cancer pain, impairing daily and social activities, and carrying a significant economic burden measurable in billions of dollars annually [93, 94]. Estimations are that there are more than 4.7 million THA operations done annually in the whole world, with significant portion due to DDH [95]. Average total expenses of THA treatment are about 20,000 euros per patient [17], with great variance. For illustration, in Serbian healthcare system, it is less than 10,000 euros using the same modern implants.

In accordance to non-maleficent approach, detailed patient examination and utilisation of all diagnostic and therapeutic procedures that are indicated for suspected condition in every patient, always leads to better clinical outcome. But in everyday practice, there is usually more than one option for every step in patient management. These options sometimes differ not only in side effects, reliability, safety or indication requirements but also in technical and financial availability. That's where statistics and economics come to interfere with strictly medical issues. In some cases, such as DDH, many factors need to be considered in order to see the whole picture [96–98].

For instance, introduction of ultrasound examination to clinical screening for hip dysplasia revealed that some clinically positive cases are false positive, but also vice versa; it brought a fraction of clinically normal, but sonographically dysplastic or lax hips. Since it incurred extra cost and organisational effort, subsequent justification had to come from economic studies [45, 68].

Factors that contribute to late diagnosing of DDH include inconsistent implementation of screening protocol, lack of appropriate awareness of the disease and its complications and insufficient training in proper and timely detection of DDH and therapeutic actions [42, 99]. Important but often neglected issue in this situation, where operative treatment makes the method of choice,

is the involvement of parents (usually mother) in the process of treatment. Mothers stay with their child for the whole duration of the treatment—usually for 3–6 months following open reduction, with the child immobilised in bed by plaster cast or skin traction, in hospital and/or at home for weeks. This is tangled with many new problems involving the patient itself (such as feeding, hygiene in the cast, dressing, sleeping, transport), mother (employment status, social isolation, existing physical and mental health condition) and the rest of the family (altered daily activities) for a long period of time [100]. There is a general lack of information and outpatient support about recovering child's complex needs during that period [101, 102].

Summary

There is considerable diversity in opinions worldwide regarding both diagnostic and therapeutic approach to DDH. Besides orthopaedic, many other factors could contribute to it: demographic, socioeconomic and differences regarding healthcare organisation [5]. Reflecting this diversity, in this book authors will present their experience and opinion on several important issues regarding DDH: screening for DDH, biomechanical considerations, diagnostic procedures in all age groups, treatment modalities of hip dysplasia and dislocation in childhood, and dealing with the consequences in adulthood.

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