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Congenital hip dysplasia: The importance of early screening and treatment

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Abstract

Congenital Hip Dysplasia (CHD) is characterized by a hip joint dislocation between the femoral head and the acetabulum, with a multifactorial etiology. This disorder can be an isolated condition or the manifestation of a syndromic condition, and it has been estimated with higher rates than registered, with a predominance in female sex and left side; risk factors are now defined. In Italy, the incidence rate is 3-4%, with significant regional differences: higher in Lombardy and lower in Sicily. Because clinical examination alone is insufficient to diagnose CHD, it is supplemented with ultrasonography and X-ray if necessary. Surveillance, static or dynamic splints, or osteotomies are the only treatment options. The goal of this study was to evaluate our experience in terms of management and conservative treatment of all newborns from January 2018 to May 2022: female sex and left hip were major involved, risk factors were not significant in our case, but results from early diagnosis and treatments, in terms of better outcome, were interesting. After a strict 6-month follow-up period, 89.13% of the patients were classified as grade Ia or Ib according to the Graf classification system. Finally, we emphasize the importance of early universal screening and subsequent diagnosis to allow for early treatment of the disorder, at an age when conservative treatments can yield good results.

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Introduction

Congenital Hip Dysplasia is a developmental disorder defined by anatomical aberrations that involved building structures of the hip joint, such as abnormal acetabular cartilage and subsequent laxity of the capsule. It prejudices the fit of femoral head in hip joint, leading to different functional disorders depending on anatomic aspects: from mild laxity to complete dislocation of the joint, avascular necrosis of femoral head, early osteoarthritis, secondary femur damage and movement problems if not early treated. Complications are typically characterized by the need of performing a total hip arthroplasty in young adults.[1]

In literature is also used the Anglo-Saxon term "Development Dysplasia of the Hip (DDH)", focused on its evolutionary potential during lifetime and the related pathologic features; we prefer referring to the aforementioned disorder using "Congenital Hip Dysplasia (CHD)" in accordance with Italian Pediatric Association of Orthopedics and Traumatology (SITOP) to better convey its association with genetic/epigenetic aspects and its natural history as a dynamic condition which may deteriorate or improve lifetime.[2]

CHD can be an isolated condition or associated with genetic neuromuscular disorders as a syndromic condition, such as arthrogryposis, myelomeningocele, Ehlers-Danlos and Larsen's syndrome. In these cases, clinical features are evident and help us in diagnosis. Recent association studies in literature tend to consider CHD as a multifactorial disease including genetic, epigenetic and environmental risk factors at the base. Genetic factors are multiple pathway-related genes; as epigenetic factors we can mention DNA methylation, histone modification and non-coding RNA-associat-

ed gene silencing that are involved in variation of specific gene expression for building joint's structures.[3]

Unfortunately, our knowledge is lacking, the exact etiology is still unclear and debated.

Risk factors include female sex, a positive family history (especially first-related relatives), breech presentation, prematurity, gestational age, multiple pregnancy, high weight at birth, clubfoot disorders, oligohydramnios, fetal malposition, presence of different malformations and limited hips abduction. The last 3 mentioned can lead to "packaging problems" creating a constrictive intra-uterine environment, subsequent malposition and maldevelopment of the hips.[4] CHD is one of the most frequent skeletal anomalies and world-wide spread but with different prevalence: high in Asia and Europe, especially Mongolian, Mediterranean and North-European population; low in Sud-Africa and Sub-Saharan area; decreasing in Japan. Many Authors sustain a connection between infants swaddling and DDH, where legs are extended, and hips are in extension and adduction position.[5] In Italy the average incidence is 3-4%, with higher rates in Emilia Romagna (5%), Val d'Aosta, Marche, Basilicata (4%), and Lombardy, where it can reach the 15-20% of the population, contrariwise lower rates are registered in Sicily (2.5%).[6] Real rates are estimated to be higher than registered reports, depending on the effectiveness of early screening and diagnosis. CHD may be one of the major causes for a Total Hip Arthroplasty (THA) in young adults as R.I.P.O. shows: it stands at 9%, after primary coxarthrosis (69.4%) and fractures of femoral neck (9.2%).[7] Dysplastic hip(s) may be observed within clinical examination in the presence of following features: lower-limb length discrepancy, asymmetric thigh or gluteal folds, painful or, on the other hand, asymptomatic motion; limited motion and asymmetric abduction are assessed by specific tests, the most performed ones are the Ortolani and Barlow tests.[8-9] We can consider Barlow's test positive when hip dislocates, Ortolani's test when hip relocates with evidencing palpable sensation of the femoral head moving into the acetabulum and a clunk (Snap Mark). Specificity and sensitivity of these tests are examiner-dependent: experience and training have an important impact on positive predictive values; they still can identify CHD in 54% of cases.

Gold standard for diagnosis is the ultrasonography, which allows the visualization of femoral head position and the anatomic relation between femoral head and acetabulum, specific parameters such as acetabular depth and inclination, static or dynamic evaluation. After 4-6 months of age, the femoral head ossification nucleus is radiographically visible, and it can be performed to manage severe CHD or to prove the effectiveness of the adopted treatment; fundamental radiograph signs are: Perkins line and arc of Shenton.[10] CHD can be treated in different modalities, depending on the severity of the dislocation. Current guidelines suggest for a early diagnosis and treatment if necessary, based on the knowledge that after six months resolution is very unlikely.[11] Early management can be either represent by strict surveillance, including clinical examination and ultrasonography or static/dynamic harnesses to be worn most of the time. Static splints are less used nowadays because they promote a "rigid reduction" and seem to have a higher rate of complications, first of all avascular necrosis of the femoral head. The most accepted indication is in unstable hip that need to be centered. On the other side dynamic splints promote a "dynamic reduction" of the hip, leaving the child free to move within a permitted range and maintaining the hip in flexion and abduction; they are valid therapeutic options for CHD, especially if applied early (within 6 months). Besides less complications are recorded in systematic reviews published in literature.[12] For severe and/or irreducible hip dislocation the only valid options are reduction undergo general anesthesia and/or surgical treatment, such as pelvic/femoral osteotomy and periacetabular osteotomy, to prevent worsening of clinical features with degenerative joint disease, postural scoliosis, pain and gradual disability.[13] Objective of this study was evaluating our experience in terms of management and conservative treatment of all newborns with diagnosis of congenital hip dysplasia from January 2018 to May 2022, in conjunction with Department of Pediatrics and Radiology A.O.U. Sassari. Here we report the analysis of all collected data, in order to discuss and compare our results to other studies in literature.

Materials and Methods

Neonatologists or Pediatricians with experience were involved in clinical assessment based on the evaluation of hip stability, using Barlow and Ortolani tests and noting potential limits in abduction.

Orthopedics managed conservative treatment and subsequently follow-up in conjunction with Neonatology and Pediatric Units, as a shared protocol; Radiology Department were involved whenever necessary. According to the screening program for CHD that has been recently adopted in Italy, all newborns had a clinical hip examination within the first days after birth and before being discharged from Neonatology Department. A hip ultrasonography evaluation was performed within the first 6 weeks after birth, using ESAOTE machine with 7.5 MHz linear probe and adopting Graf technique (Table 1) to assess hip morphology and stability.

In case of suggestive clinical signs and/or positive ultrasonography for hip instability, dislocation and/or in the suspicion of CHD, all newborns occurred to our attention for a specialistic examination. In our dedicated Clinic Room, we review the ultrasonography exam and performed a physical assessment, including Barlow and Ortolani tests. Pertinent risk factors were reported from medical history in patient's record, in particular: family history (first-degree relatives), breech presentation at delivery, prematurity, multiple pregnancy, oligohydramnios, fetal malposition, presence of different malformations and limited hips abduction.

From January 1st 2018 to May 31th 2022 we registered 46 patients: of these, 23 infants had CHD, for a total of 69 dysplastic hips; depending on the severity of clinical features, they were treated in a conservative way with specific harnesses or splints and, subsequently, evaluated every month in follow-up until the 6th month after birth. Severe detected cases underwent to close clinical monitoring and radiological pelvic exams, at the beginning and when the baby learned to walk.

Results

Inclusion criteria for patients with CHD diagnosis were as follows: i) age less than 3 months at the time of diagnosis, ii) ultrasound exams reporting Graf grade of dysplasia during the study period until significant improvements or its echo graphic normalization, iii) clinical data, iv) a minimum follow-up at least of 6 months, v) non-surgical management and early treatment.

Exclusion criteria were as follow: i) age more than 3 months at the time of diagnosis, ii) the presence of systemic and neuromuscular syndromes.

The average follow-up time was 6 months.

The study included 46 patients, of these we counted 37 females and 9 males with a male-female ratio quoted as 4:1 approximatively; moreover 23 had a single hip dysplasia (50%) and 23 had bilat-

eral hip dysplasia (50%); overall we managed and treated 69 dysplastic hips (Figure 1 and 2).

The main side involved was the left (38 in total, 55.1%), even the worst ones in terms of clinical and radiological features.

We graded the severity of CHD as Graf classification system; in a total amount of 69th we registered: 35 patients as grade IIa+ (50.72%), 14 as grade IIa- (20.29%), 4 as grade IIb (5.80%), 8 patients as grade IIc (11.59%), 5 patients as grade IId (7.25%), 3 patients as grade IIIa (4.35%).

Regarding risk factors we noted that 9 of all patients had a breech presentation at delivery (19.57%), 13 had positive family history for hip dysplasia (28.26%), 1 newborn from a multiple birth (2.17%), 3 with other congenital malformation associated (6.52%).

Clinical examination showed 3 significant Ortolani maneuvers, 11 cases of limitation in abduction motion of the hip instead other 5 cases of hyper-adduction.

We performed a clinical and radiological follow-up, using ultrasonography and pelvic radiography; X-rays became necessary in 16 cases (23.19%) of mild-severe CHD (Graf grade > IIb) and it was performed when babies started to walk by themselves.

Non-surgical treatments or observational follow-up were applied to all 46th patients (Table 2), following actual acknowledged guidelines.

In conclusion, 3 newborns did not complete all their care pathway, 2 newborns needed to use dynamic splinter, 2 infants had irreducible dysplasia and they were directed towards Specialized Hospitals for performing pelvic osteotomy.

Outcomes of our management were judged as successful (41 out of 46 cases, 89.13%) when hip achieved concentric and stable reduction classified as Ia or Ib Graf grade, with or without using abduction dynamic splints (Tubingen, Ferrara, Pavlik).

Table 1. Graf grades are schematically described in this Table; this classification system based on ultrasound evaluation of specific landmarks (angle α and angle β), permits to classify newborns' hip(s) in term of maturity and stability. Data taken from Jacobino *et al.*[14] Andreacchio *et al.*[21]

Graf Grade	Description	Angle α	Angle β
I	Mature hip	> 60°	**Ia** → β ≤55° **Ib** → β >55°
II	Immature hip **IIa+** → 0-12 weeks, adeguate bony roof **IIa-** → 6-12 weeks, deficient bony roof **IIb** → over 12 weeks, deficient bony roof	50°-59°	>55°
	Critical hip **IIc** at any age	43°-49°	≤77°
	Decentring hip **IId** at any age	43°-49°	>77°
III	Dislocated hip, based on cartilage roof. **IIIa** → normal echogenicity **IIIb** → hyperechoic	<43°	>77°
IV	Dislocated hip	<43°	Any

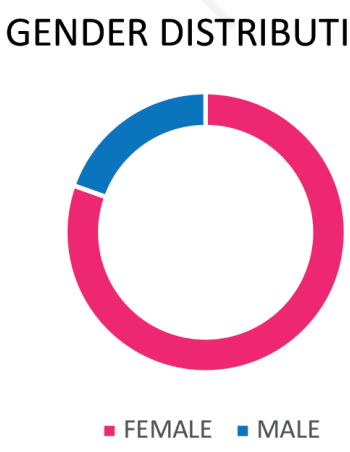

Figure 1. The figure represents the gender distribution that we registered in our study: 37 female (80.43%) and 9 males (19.57), with a female-male ratio quoted as 4:1 approximately.

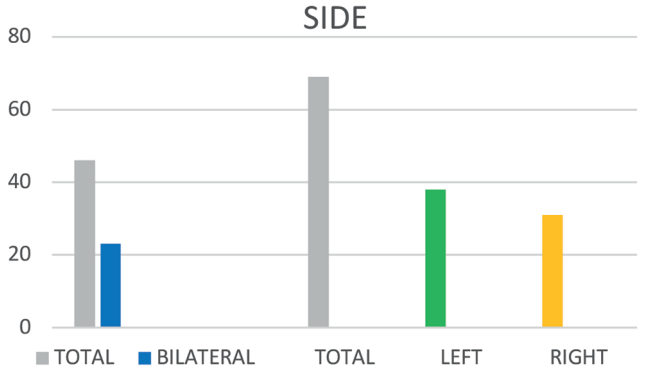

Figure 2. In our data, we collected 23 patients affected by single-side hip dysplasia (50%) and 23 patients with bilateral hip dysplasia (50%); we managed and treated a total of 69 dysplastic hips. Of these, left side was involved in 38 cases (55.1%) and right side in 31 cases (44.9%): it is showed in the second half of the graph.

Table 2. Report of conservative treatment applied to patients in our study, a total of 46 children with CHD diagnosis: 40 cases were treated with dynamic splints (Ferrara, Pavlick, Tubingen), 1 patient worn a specific panty and 5 newborns underwent to close observational follow-up only.

FERRARA	PAVLICK	TUBINGEN	PANTY	FOLLOW-UP
18 (39.13%)	8 (17.39%)	14 (30.43%)	1 (2.17%)	5 (10.87%)

The correspondent percentages are reported in brackets.

Discussion

We showed our results, in connection with data that we can find in literature: there were mostly affected female sex and left hips; moreover, in bilateral CHD, left side was the worst one in terms of Graf grade.[1-13]

Clinical examination itself was not decisive in the majority of cases, instead ultrasonography was the gold standard. Hip ultrasonography can identify the presence of hip dislocation and other anatomical markers, leading us to classify each patient in different range of severity, as Graf defined through the measurement of angle between landmarks, alpha and beta; in our study case the most frequent type of CHD was IIa+.

We confirmed the importance to extend this investigation in suspicious CHD or as early screening test. [14] We firmly sustain the basic role of ultrasound exam, and it was demonstrated by the analysis of risk factors: we registered breech presentations at delivery and positive family history in our data set, but the percentage values were not significant. For this reason, we cannot agree with Authors who regard universal ultrasound screening unnecessary and dispensable.[15] Otherwise, we considered a proper point of view the position of SITOP, proposing universal ultrasound screening instead of selective ones.[16,17]

The importance of an early screening and subsequent diagnosis of CHD is also remarkable by outcomes: we observed consistent improvements of the abnormal condition with resolution of the disorder in most of cases; our study included newborns who underwent prior examination less than 3 months age. In only 2 patients there was the necessity to lead them to Specialized Hospital for concluding the health treatment.

Thus, we can treat and manage patients in a conservative way without applying surgical approaches which are still unavoidable in severe CHD and DDH.[18,19]

We preferred dynamic splints in presence of pertinent indications and whenever the conditions of hip dislocation permitted to handle it in that way; we obtained short terms results withing follow-up surveillance, as we could reach from upper grades to Ia or Ib as a final evaluation.[20] The surveillance by itself was not effective, indeed 2 patients were subsequently treated with dynamic splints.

We are aware about the fact that our casuistry is not statistically significant, but it has been useful to evaluate our management and consequent results: we are satisfied for our success in terms of patients' clinical and radiological improvements.

Conclusions

Indeed, we would like to remark the importance of universal screening, early diagnosis and treatment in terms of obtaining better outcomes within the first months after birth, which can be resolutive for congenital hip dysplasia.[21]

Our suggestion for the future is create a mid-long term register of patients to record any pertinent and clinical changes regarding this pathology that remains unclear and an interesting disorder for literature studies, where distinguished Authors debate on its management and treatment.

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