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Pathogeny and natural history of congenital dislocation of the hip☆

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ABSTRACT

Based on a review of the literature, the authors have made a critical study of several etiological factors. Endogenous factors such as acetabular dysplasia, increased anteversion of the femoral neck, and capsular laxity support the genetic theory but are neither constant nor necessary and are only facilitating factors. The major factor seems to be a mechanical one linked to the position in the uterus: hyperflexion with adduction and external rotation *constituting the dislocating foetal posture combined with* abnormal pressure on the greater trochanter and leading to expulsion of the head upward and backward. This theory can explain the natural history of C D H which is first, at birth a hip instability followed by two possible evolutions: either persistent luxation becoming irreducible or spontaneous stabilisation leading sometimes to complete healing or to residual abnormalities (subluxation or dysplasia). This concept suggests practical conclusions: the importance of an early diagnosis, the selection of the signs of the hip at risk, the pattern of prevention, the role for non-clinical investigations, the principles of the treatment based on postures, the indications for the different types of treatment.

Although many factors have been incriminatedin the aetiology of congenital hip subluxation, dislocation, and dysplasia, none of the pathogenic hypotheses is fully satisfactory. The confusion resulting from this uncertainty generates a number of issues:

- terminological issues, since the classic term "congenital dislocation of the hip" (CDH) is widely considered to be inappropriate and is often replaced by "dislocating malformation", "dislocating dysplasia" or, more recently, 'developmental displacement of the hip' (DDH)[Klisic [30]];
- diagnostic issues, as the meaning of the clinical instability remains controversial and no consensus exists about the role for radiography and, above all, for ultrasonography [Bonnard [5], Graf [25], Téot and Deschamps [57,58]];
- public health issues, because depending on the pathogenic concept the strategy should focus either on prevention [Klisic [29,30]] or on screening;
- and treatment issues, since the natural history of CDH largely dictates the therapeutic indications. The main concern is the risk of overtreatment that places a costly burden on society and can induce iatrogenic complications (avascular necrosis of the femoral head). In addition, improved knowledge of the cause of the dislocation facilitates the conduct of the treatment by providing a sound underlying rationale, thereby diminishing the reduction failure rate.

Based on published work by Le Damany [34], Faber [21], Ortolani [37], Salter [43], and Tachdjian [56], CDH is usually viewed as a hip development abnormality that starts in utero and becomes apparent at birth or within the first few post-natal months (Table 1).

As clearly demonstrated by Gardner [23], true dislocation cannot occur in the embryo before the formation of the articular cleft. Clavert [12] administered antimitotic agents to pregnant rabbits during the period of hip embryogenesis and obtained a marked increase in the prevalence of CDH, suggesting that CDH might be due to an overall hypoplastic malformation of the hip region. This hypothesis is not confirmed, however, by everyday clinical practice.

Davies [16], Dyson [20], Ilfeld [28], and Tredwell [61] concurred with Wynne Davies [66] that CDH may exist as two distinct aetiological forms: a form due to joint hyperlaxity that can be detected at birth based on perception of a positive Ortolani manoeuvre and a form related to subluxating acetabular dysplasia, which escapes detection by neonatal screening and is diagnosed only at a later age.

However, our experience with neonatal CDH screening and efforts to identify clinical signs that are more subtle than the palpable clunk during the Ortolani manoeuvre [44]; our anatomic [45], radiographic [46–48] and, more recently, ultrasonographic [49] studies; and a critical review of the literature lead us to suggest a unifying theory of the pathogenesis of CDH.

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Table 1
Classical view of the natural history of CDH (from Salter).

In utero	Shallow acetabulum Femoral anteversion	*Dysplasia* ↓
Birth	Hip no longer flexed Joint laxity	*Dislocatable hip* ↓
After birth	Gradual hip extension Tight swaddling	*Dislocation*

1. Study of the various aetiological factors

The aetiological factors fall into two categories, endogenous or constitutional factors and exogenous or mechanical factors.

1.1. Endogenous factors

These consist in a primary hip abnormality that may involve the acetabulum, femur, or joint capsule. The existence of endogenous factors suggests a role for genetic susceptibility, which may explain the greater frequency of CDH in girls (5 girls for 1 boy) and in certain geographic areas, ethnic groups, and families (in 3% to 12% of cases). When one twin has CDH, the frequency of CDH in the other twin is 40% for monozygotic twins and 3% for dizygotic twins [Carter [8], Idelberger [27]].

Thus, although genetic factors cannot fully explain the occurrence of CDH, they play an undeniable role [Fuhrmann [22]]. The genetics of CDH in France have been thoroughly investigated by Le Marec and Roussey [35], who ruled out monogenic inheritance but also found that a multifactorial theory was unsatisfactory. Nevertheless, they indicated that the table developed by Stalder [54], which implies a multifactorial pathogenic process, is an easy means of estimating the genetic risk: about 10% in siblings of a boy with CDH and 3% in those of a girl with CDH, and about 5% in the first-born child when one of the parents has CDH.

1.1.1. Acetabular dysplasia

The hip dysplasia theory advocated by Faber [21], Courtois [15], and Klisic [30] long prevailed. This theory constitutes the rationale for the ultrasound studies conducted by Graf [25]. Nevertheless, many lines of evidence argue against it.

- Experimental studies in animals during the growth period have established that hip dislocation induced by abnormal lower limb position [Asplund [2], Michelsson [36], Salter [42], and Yamamuro [52]] or by damage to the connecting structures of the hip joint [Langenskiold [32], Sibrandj [50] and Smith [52]] cause secondary acetabular deformities. The acetabular is distorted into an oval shape, and wear creates a dislocation groove located either in the antero-superior sector if the hip was extended (Salter) or in the postero-superior sector if the hip was left in flexion (other experiments). The acetabular dysplasia is reversible if the femoral head is returned to its normal position in the cavity. In all these experiments, the dysplasia was induced by the dislocation, contraindicating the theory that dysplasia causes the dislocation.
- In studies on the natural history of hip dysplasia without dislocation in infants [Coleman [13], Geiser [24], Pratt [38], Seringe and Hass [47]], none of the patients progressed to subluxation or gradual dislocation (Fig. 1).
- Comparisons of the outcomes of treated and un-treated hips [Seringeand Haas [47]] established a distinction between secondary dysplasia (due to the dislocation), which was reversible; and primary dysplasia, which was not influenced by the treatment (Fig. 2).

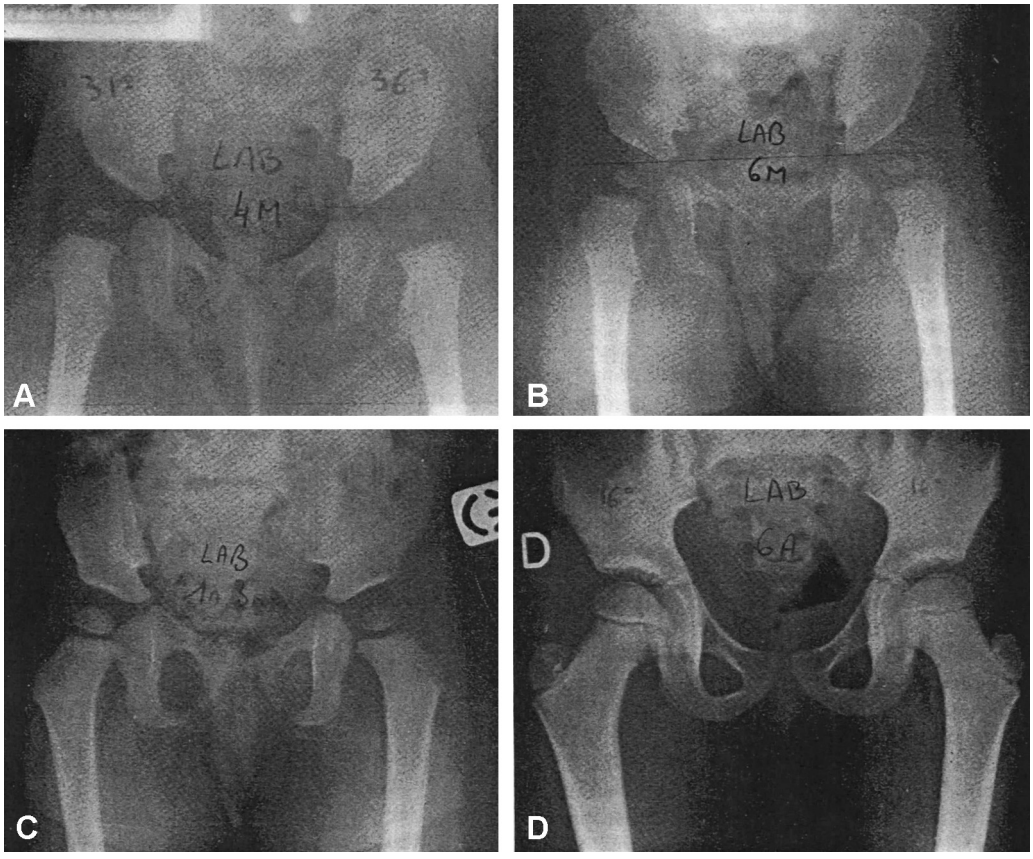

Fig. 1. *Natural history.* (A) *At 4 months of age*, bilateral acetabular dysplasia is visible and the proper centring of the femoral heads is in doubt (in part due to the external rotation of the femurs). (B) *Appearance at 6 months of age*. (C) Continued spontaneous improvement at *15 months of age*. (D) The hips are normal at *6 years of age*.