PRIMARY PROTRUSIO ACETABULI
Report of an Affected Family

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Since its initial description in a dissection specimen by Otto in 1824, protrusio acetabuli has provoked much discussion on etiology and pathogenesis. It seems now generally accepted that a primary, so-called idiopathic, type does occur, and the object of this paper is to report a family in which familial factors, probably genetic, appear to play an important part.

Apart from a post-mortem specimen recorded by White (1883), the literature on the subject remained almost entirely Continental until Hertzler (1922) reported four cases in America. His title “osteo-arthritic protrusion of the acetabulum” was taken from the German “intrapelvine Pfannenvorwölbung” (Valentin and Müller 1921). Verrall (1929), obviously unaware of previous reports, described a single bilateral case under the heading “arthrokatodysis”, meaning literally subsidence of a joint. Pomeranz (1932) reviewed the seventy-nine cases published in the 100 years or more since Otto’s original description, but made no mention of a primary or idiopathic form. In 1934 Golding simplified the classification into the three currently recognised

<table>
<thead>
<tr>
<th>Type of protrusion</th>
<th>Articular cartilage</th>
<th>Acetabular floor</th>
<th>Head of femur</th>
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<tbody>
<tr>
<td>Primary</td>
<td>Normal width</td>
<td>Very thin</td>
<td>Unaltered</td>
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<tr>
<td>Arthritic</td>
<td>Absent or thin</td>
<td>Thickened by new bone accretion</td>
<td>Sclerosed, irregular, pointed</td>
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<tr>
<td>Secondary</td>
<td>Various types of destructive bone disease</td>
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Fig. 1
Pedigree of the family. The roman numerals indicate generations. The arabic numerals indicate age in years. The capital letters indicate the initials of the members.
groups (Table I). The arthritic group is now commonly accepted as the late stage of the primary group (Rechtman 1936, Gilmour 1939, Brailsford 1953, Friedenberg 1963).

As early as 1903 Eppinger had postulated that the condition was caused by delayed ossification of the Y-shaped cartilage of the acetabulum during adolescence. Despite the introduction of radiological diagnosis by Scherlin (1911), his views were almost universally ignored until Schaap (1934) on clinical grounds and Golding (1934) on the radiographic appearances first clearly segregated a primary (idiopathic) type from the heterogeneous group of secondary causes. Schaap pointed out that this type affected mainly females, that it was essentially bilateral and that evidence of inflammation was lacking. He felt, as did Overgaard (1935) and Rechtman (1936), that a congenitally too deep acetabulum initiated the process. However, Gilmour (1939) noted that there was often a disturbed pattern of adolescence in girls and that this was probably associated with premature fusion of the Y-shaped cartilage, leading to the development of a deep acetabulum. In this regard Morton and Hayden (1941) pointed out that in the pre-pubertal child there was already a normal inward bulge of the acetabular region, which remodelled with subsequent growth. Alexander (1965) agreed with Gilmour and postulated that it was failure of this remodelling that led to the development of a deep acetabulum. He maintained that the earliest sign of this failure was persistence of the “beaking” of the Y-cartilage as seen in the antero-posterior radiograph.

Rechtman (1936) seems to have been the first to suggest that the condition might be familial. In three of his five cases there were familial features. Gickler (1937) reported an affected brother and sister, and Brentrup (1942) two mother and daughter pairs. Racial differences have been noted by at least two authors. Friedenberg (1953), reporting seven female cases, noted that six patients were negroid. Crichton and Curlewis (1962) reported an extraordinary incidence of 25.7 per cent in 226 pregnant Bantu women, compared with 5.7 per cent in Indian and 2.9 per cent in European pregnancies. Solomon (1967) noted an overall incidence in the Bantu of about 11 per cent. Francis (1959) alone seems to have instituted a familial investigation along classical Mendelian lines. In a study of six family trees, one family showed a striking hereditary influence through three generations, probably from a dominant gene with incomplete penetrance.

**FAMILY CASE REPORT**

The pedigree of this family is shown in Figure 1. The proband (J. D.) was a man aged thirty-nine years who presented with pain and stiffness of both hips, increasing over a number of years (Fig. 5). He was of low mentality (though literate) and unmarried. His mother (C. D.)
FIG. 3
Generation II, A. W., aged 60 years. Alert woman, the elder sister of the proband, and the only procreative member of the generation. Long-standing pain and stiffness both hips, severe on the left.

FIG. 4
Generation II, R. D., aged 54 years. Elder brother of the proband, and of low mentality. Severe pain and stiffness both hips of long duration.

FIG. 5
Generation II, J. D., aged 39 years. The proband. A man of low mentality, and with severe stiffness of both hips of long duration.
Generation III, C. T., aged 37 years. A woman with deep acetabuli and admitting to occasional nocturnal pain in the right hip. Three daughters (Figs. 9 to 11).

Generation III, C. W., aged 34 years. A man with a deep right acetabulum, symptomless. Unmarried.

Generation III, A. H., aged 23 years. A woman with deep acetabuli but without symptoms—one son aged 3.
(Fig. 2) was a sprightly woman of eighty-three with no arthritic disability of the hips. The elder brother (R. D.) (Fig. 4) was aged fifty-four. He gave a similar, though longer, history to that of J. D., culminating in fusion of one hip two years previously. He was of low mentality and unmarried. The elder sister (A. W.) (Fig. 3) was a normal married woman aged sixty. She had never sought treatment but had suffered increasing stiffness of the hips for years, with recently increasing pain on the left side.

A nephew (C. W.), son of A. W. (Fig. 7), was aged thirty-four years, unmarried and normal. There were two nieces (daughters of A. W.) and their children. The elder niece, C. T. (Fig. 6), aged thirty-seven, was a normal married woman but admitted on questioning to slight stiffness and nocturnal aching in the right hip. Her daughters, J. T., aged fourteen years (Fig. 9), A. T., aged twelve years (Fig. 10), and K. T., aged six years (Fig. 11), were normal. The younger niece, A. H. (Fig. 8), aged twenty-three years, was normal, as was her son D. H., aged three years.

A summary of the radiographic and clinical features of each member is given in Table II.

**DISCUSSION**

Marked inward protrusion of the acetabulum is immediately obvious, but there is no clear definition of the earliest degrees of protrusion, and even less certitude surrounds the definition of a deep acetabulum. Pomeranz (1932) and Overgaard (1935) implied that protrusion was not present until the pelvic floor of the acetabulum intruded beyond the linea
终端is by a few millimetres or more. Schaap (1934) regarded an “abnormally deep” acetabulum as an earlier stage, but gave no criteria for measuring this. Overgaard (1935) introduced the use of Köhler’s (1928) lines, the so-called “tear-drop” lines. Crossing of these lines in a well centred antero-posterior film constituted the earliest degree of a “deep acetabulum”. Friedenberg (1953) in addition employed the C/E angle of Wiberg (1939) but gave no precise figure for the upper (deep) limit of normal.

In assessing the family reported here great difficulty was experienced in establishing objective criteria of normal or abnormal acetabular depth, but much less difficulty in deciding subjectively on casual inspection of the radiographs. Arbitrarily a C/E angle of 45 degrees (+5 degrees), coupled with a crossed tear drop, has been selected as constituting the beginning of abnormal depth. C/E values much over 50 are easily picked as “deep” whereas below 40 the hip is clearly normal. On this basis all recorded members of generations I to III had deep or protruded acetabuli.

Of the children in generation IV, J. T., aged fourteen years (Fig. 9), was judged to have deep acetabuli. A. T., aged twelve years (Fig. 10), showed persistence of the inward bulging described by Morton and Hayden (1941) and of the “beaking” described by Alexander (1965): her hips were judged to be normal. Neither case appeared too advanced for remodelling to occur. The Y-cartilages in the hips of both girls were fusing or had fused, but fusion did not appear premature on the figures given by Schinz, Baensch, Friedl and Uehlinger (1951). The child K. T., aged six years (Fig. 11), showed normal Y-cartilage beaking for her age.

Although the family tree is incomplete in regard to the husbands of the first three generations, it seems probable that there is an expression of a dominant gene, or polygenic
complex, resulting in abnormally deep acetabuli throughout four generations and with gross primary protrusio acetabuli in all three members of the second generation.

**SUMMARY**

1. The literature of primary or idiopathic protrusio acetabuli is reviewed with particular reference to familial and racial influence on pathogenesis.
2. The radiological criteria of a “deep” acetabulum and of a “protruded” acetabulum are discussed.
3. Four generations of a family are presented in which all three members of the second generation showed marked protrusio acetabuli. In the remaining generations most members appeared to have abnormally deep acetabuli.
4. It is concluded that this family shows a strong familial tendency to deep or intruded acetabuli. The family tree, though incomplete, suggests a genetic influence of a dominant type.

**REFERENCES**


