BILATERAL GLENOID HYPOPLASIA

Report of Five Cases

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The five patients under consideration, all of whom were seen within three years, had in common a bilateral defect of the glenoid articular surface of the scapula accompanied by flattening of the humeral head. In some there were associated abnormalities in the axial skeleton. Disability was marked in only two cases.

Reports of only a few similar cases have been found in the literature. Giorgio (1927) and Heupke (1928) reported "glenoid dysplasia and clavicular spurs." Brailsford (1944) mentioned those and described three further patients (two women and one girl of nine years), all with bilateral defects of the glenoid, webbing of the axillae and limitation of abduction; he regarded them as congenital deformities, and mentioned the possibility of concomitant spurring of the clavicles and tendency to humerus varus. Scaglietti (1938) and Lucas and Gill (1947) discussed cases of humerus varus with secondary glenoid hypoplasia. Their cases were ascribed to birth injuries with epiphysial separation and later moulding. Scaglietti noted that bilateral defects occurred after breech delivery, whereas unilateral deformity usually followed normal vertex birth. Andesin (1948) described congenital defects of the humeral heads in two adults. His cases were similar to the cases described here, but the defect of the head was much more marked and the glenoid surface was decidedly convex.

CASE REPORTS

Case 1. Schoolboy aged fifteen years. He stated that he had never been able to lift his arms above shoulder level. There was no history of maternal illness in pregnancy, birth was normal, no relevant childhood illness or injury was known and no other member of the family was similarly affected. Clinically, apart from moderate underdevelopment of the deltoid and pectoral muscles, the outward appearance was normal. Shoulder movements, however, showed abduction in the scapular plane to only 90 degrees, even with the arms in lateral rotation (Fig. 1). The thoraco-scapular movement was normal, the restriction of abduction being entirely in the scapulo-humeral joint. Rotation was free. Hip and cervical spine movements were full. No other congenital abnormalities were noted.

Fig. 1
Case 1—Showing shoulders abducted to fullest extent.
Radiographic examination—On each side there was a defect of the inferior angle of the articular surface of the glenoid (Figs. 2 to 5). The superior epiphysis was present but ununited. There was slight flattening of the medial part of the upper humeral epiphysis. The lateral end of each clavicle showed a curious hooked appearance and a bony "knob" protruded from its inferior aspect at the site of insertion of the conoid and trapezoid ligaments (cf. Brailsford 1944). The coracoid processes were large and prominent. No abnormality was detected in the cervical spine or pelvis.

Case 1—Right and left shoulders with arms by the side.

Case 1—Right and left shoulders with arms abducted to fullest extent.

Case 2—Man aged twenty years. Complained of inability to raise either arm above shoulder level from infancy. He was keen on farming but the disability compelled him to take up an office job. When seven months pregnant the mother had suffered from protracted vomiting. Delivery was at full term. The forceps was used for a normal vertex presentation. There were no childhood incidents and the family history was negative. In general the patient was well developed, but stood with a moderate thoracic kyphosis and slight elevation of the left shoulder. There was also a.
"funnel chest" deformity. Shoulder movements—Rotation and forward flexion were normal. Abduction both in medial and in lateral rotation was markedly limited (Fig. 8). When abduction was attempted the humeral heads seemed to subluxate backwards on the glenoid. Rotation at the left hip was slightly limited but there was no obvious asymmetry of the lower limbs.

Radiographic examination—On each side there was hypoplasia of the glenoid fossa affecting mostly its inferior angle; the articular surface was almost vertical and merged into the axillary border of the scapula (Fig. 7). In the axial view the posterior part of the glenoid especially was poorly developed (Fig. 8) (hence the clinical finding of backward shift of the humeral heads in abduction). The coracoid process was large, and the articular surface of the humeral head was flattened. The only associated abnormalities detected were wedging of the C7 vertebral body and spina bifida of the T1 vertebra. The hips appeared normal.

Case 3 Man aged forty years. Joiner. He had noticed no disability of the limbs except that from childhood the left arm had been slightly weaker than the right. He was first seen as a casualty after sustaining a fracture of the left glenoid fossa in a fall, and it was thus that his abnormal

Fig. 6
Case 2 Showing shoulders abducted to fullest extent.

Fig. 7
Case 2 Left shoulder seen in antero-posterior projection (Fig. 7) and in axial view (Fig. 8).
The right shoulder was similar.

Fig. 8
limbs were discovered. Clinically there was obvious hypoplasia of the left arm and shoulder girdle. The left lower limb was three-quarters of an inch shorter than the right owing to congenitally short femur. There was asymmetry of the chest from flattening of the thoracic cage on the left. Movements of the right shoulder were normal, although those of the left shoulder were restricted at that time because of the injury, later examination showed practically a full range of abduction. There were no neurological signs.

**Radiographic examination**—On each side there was a defect of the glenoid fossa affecting predominantly the inferior half of the articular surface, as in the previous cases (Figs. 9 and 10). (There was a recent fracture on the left side.) Each humeral head was of oval shape. There was a small bony knob on the inferior aspect of each clavicle like those described in Case 1. There were bilateral cervical ribs, but no congenital abnormality was noted in the thoracic cage or hip joints.

**Case 4**—Woman aged forty-five years. Had never had trouble with her shoulders until her first attendance two years ago. At that time she was suffering from mild periarthritis of the left shoulder as the result of a fall. When examined recently recovery from this was complete. She had been told that she had been born "seven weeks too soon," but otherwise pregnancy and labour had been normal. Her mother had suffered from "a bad hip" since childhood; otherwise the family history was clear. On examination the patient was asthenic but no obvious locomotor defect was present. The extremes of shoulder abduction and medial rotation, however, were limited on both sides.

**Radiographic examination**—In each shoulder there was hypoplasia of the glenoid articular surface (Fig. 11) like that found in Case 2. The humeral heads were only slightly abnormal. There was a cervical rib on the left side, and on the right an ossicle was noted near the transverse process of the C.7 vertebra.
Case 5— Man aged forty-six years. Minor. Complained of stiffness of the right shoulder after an injury ten years previously. Past history and family history were negative. Apart from moderate wasting of the right shoulder girdle and disturbed scapulo-humeral rhythm from periarthritis, physical examination was negative. The left shoulder was normal in appearance and function.

Radiographic examination—in each shoulder there was a glenoid defect like that found in the previous cases. On the right side there was slight lipping of the articular margin; otherwise the humeral heads were normal, and no clavicular spurs were noted. No radiographs of the spine or hips were available.

DISCUSSION

The cases considered here have first to be differentiated from similar acquired deformities of the shoulder. Thus Scaglietti’s (1938) description of bilateral humerus varus with epiphysial hypoplasia of the glenoid was ascribed to birth injury to the upper humeral epiphysis. There was an associated irregular sclerosis of the middle third of the clavicle with accentuation of its normal S-shaped curve. Clinically the arms were medially rotated and slightly abducted.

Other recognised causes of humerus varus produce transient softening of the humeral head in early life: rickets and Barlow’s disease are examples. Unilateral deformity may arise from injury to the epiphysis in the formative years between five and twelve, with consequent osteochondritis. Fairbank’s (1947) multiple epiphysial dysplasia can give a similar picture, but changes are present in other joints such as the hips. Erb’s palsy, producing secondary disuse atrophy of bone from muscular weakness, and the muscular dystrophies (Ashby et al. 1951) are sometimes associated with hypoplastic humeral heads and glenoid cavities.

In contrast, the present cases are considered to be congenital in nature. This view is supported by the presence in some of the cases of other skeletal abnormalities such as cervical spina bifida and hemivertebra, cervical ribs, clavicular spurs and hemiatrophy. It is unlikely that all these are coincidental. In no case is there evidence of post-natal or birth injury and no soft tissue or nervous changes were observed (the recent injuries are discounted).

Radiological studies have shown that the normal glenoid is made up of three components (Brailsford 1944). Its floor is included in the main scapular body which ossifies at approximately the eighth week of intra-uterine life. The apophysis which appears at the tenth year goes to form the upper ring of the glenoid and base of the coracoid process. Finally, the inferior segment of the glenoid is derived from a centre appearing at about thirteen years. This serves to deepen the articular surface and produce a well defined inferior articular margin. It is to be noted that the precartilage for the centre is normally already well established in utero. Rarely a separate ossicle is seen at the superior and inferior angles of the glenoid in children of about twelve years. The humeral head is formed from a medial epiphysial centre ossifying at six months, a lateral one at two years and an anterior one for the lesser tuberosity at five years.

A simple explanation of the abnormality of the glenoid in the present cases is that the precartilage of the inferior apophysis failed to develop. But this does not explain the underlying embryological processes involved. Much of the work done in this field has been on embryo chicks, the experiments involving ablation of embryonic tissue. No parallel process takes place in the developing human foetus, but certain general conclusions have been reached. These are based mainly on the work of Murray (1926) and of Fell and Canti (1934) on the development of isolated limb buds of chick embryos implanted on chorio-allantoic membrane.

1) Early joint formation is not dependent on the presence of muscle, nerve or vascular tissue. It does, however, depend on intrinsic predetermined factors—the so-called inherent mosaic. 2) Some form of joint will appear even if the presumptive joint region has been ablated (not applicable in man). 3) The shape of such a joint will depend on the amount of tissue removed or destroyed and the stage of development of the blastoma at the time of ablation. 4) In
normal joints the precartilage determines the future shape of the bone laid down. 5) In post-natal life extrinsic factors such as muscle pull or trauma influence joint shape to some extent. The full story of early bone and joint development still awaits clarification. It would therefore be presumptuous to be too categorical about the precise congenital factors involved in the causation of the shoulder defects described.

SUMMARY
1. Five cases of bilateral glenoid hypoplasia are described. Flattening of the humeral heads and sometimes other skeletal abnormalities coexisted.
2. The condition is considered to be congenital.
3. The differential diagnosis and etiology are discussed.

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REFERENCES