A CASE
OF
COMPLETE ABSENCE OF BOTH THE
UPPER LIMBS
AND OF
FAULTY DEVELOPMENT OF THE RIGHT LOWER LIMB.

BY
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(Received November 21st—Read November 28th, 1876.)

SUCH extreme congenital deformity as that exhibited in this child is so unusual that I venture to think a short account of her will not be uninteresting to the Fellows of this Society. Her singular condition attracted my attention last year when she was brought to the Middlesex Hospital to see an elder sister, then under my care in the accident ward. Observing the interest the child excited her mother allowed me to examine her and to have the accompanying photograph of her taken by Lenthall (see Plate VI). Dr. Lyall, our surgical registrar, obliged me by taking the memorandum which forms the substance of this note.

Mary B—, æt. 4½, apart from the deformities to be described, a well-grown, healthy, pretty, intelligent blonde, is the youngest of four children, none of whom, except

herself, are remarkable for any bodily peculiarity. Her father is a carman, her mother a dressmaker; both are well-formed and healthy.

The child's upper limbs are both completely absent, which gives the upper part of her naked trunk a very angulated, narrow figure. The lateral contours slope vertically, with a slight outward slant from the extreme points of the angular shoulders to a little above the level of the nipples, whence, bending inwards, they fall upon the middle ribs of a well-shaped chest. The clavicles and scapulæ appear well formed. They are of a size corresponding to her stature, and the muscles attached to them act vigorously. Below the projecting acromion no trace of humerus or of any of the distal bones of a limb can be discovered on either side; a cushion of soft tissues fills the angle between the axillary border of the shoulder-blade and the side of the chest. Below the outer end of the right collar-bone, nearly in the level of the vertical semi-diameter of the scapula, is a minute congenital scar, and slightly below and outwards from this is a little dimple. No mark of any kind is discernible on the left shoulder.

When placed upright on her left foot the right limb hangs from the trunk, its foot not reaching the ground by several inches. The thigh is very short and plump; the leg is well shaped, but smaller than the left. The foot has a moderate degree of talipes equino-varus, and the fourth and fifth toes are joined together, the fourth cleft being absent. A very short misshapen femur can be made out, and both bones of the leg are clearly present; contrasted with the femur they seem well developed. The mobility of the thigh upon the pelvis is very great; abduction and circumduction are especially free. She readily puts the thigh through most of the movements of a ball-and-socket joint. The knee-joint is loosely knit; in addition to wobbling hinge movements, the axis of the tibia can be made to rotate upon the lower end of the femur, not only in flexion, but also in extension of the knee joint, and some amount of lateral gliding is also possible.

The left lower limb is perfect. It dexterously performs many offices usually devolving on the hands. The toes are very mobile and prehensile, and so under control, that with the foot she readily grasps and steadily holds even small objects, and executes complex concerted acts, as picking up a comb and arranging her hair, feeding herself with a spoon, and even using a needle. She sometimes makes use of her right (distorted) foot to fix anything she wishes to grasp with the left.

Comparative measurements of the limbs.

		From antero-superior spina ili to malleolus internus. Inches.		From same point to inner femoral condyle. Inches.		Girth of thigh. Inches.
Left	.	14	.	7	.	11½
Right	.	8	.	—	.	12½

Her mother states that six weeks after conception she met and stopped to look at a beggar in the street who was without arms and had both feet clubbed. This did not make any impression of terror or anxiety upon her, and the circumstance had nearly passed from her memory when recalled by the inquiries of friends.

Interesting as is the vexed question of the influence of maternal emotions upon the embryo, I would suggest that the chief interest of this case lies in another direction. The support it affords to the theory that these and similar defects of the limbs are due to absence, or arrest, or misdirection of development, and not generally, as is very widely thought, to amputation of already formed limbs by their adventitious constriction. The small scars and the little appendages of the present (such as might result from the healing of a wound after removal of part or of an entire member at its junction with the trunk), and the occasional appearance of a furrow upon a limb, suggestive of its having been tightly girt by the umbilical cord or by a band of thickened membrane, are the chief evidence adduced in favour of the hypothesis of intra-uterine amputation. As regards the umbilical cord, however, it is not

easy to understand how it can become so tightly coiled around a limb as to cause this to drop off without itself being, at the same time, so stretched as would greatly lessen the volume of the umbilical arteries and vein, and so seriously prejudice the life of the embryo. Yet, as in this case, well-grown offspring are born at full term with quasi-amputated members. The scars and little appendages adduced in favour of amputation *in utero* admit of a better explanation; they may be, and probably, usually they are, vestiges of members blighted at an early stage of development from some intrinsic defect in the germ, as the author of the 'Catalogue of Monstrosities in the Hunterian Museum' and others have suggested. This is strengthened by their frequent association with other defects of a kind which cannot, from their very nature, owe their origin to any such mechanical constriction as is imagined, viz. deficiency of brain and the like, and redundancy of parts.

The total absence of scar from the child's left shoulder excludes absolutely the notion of amputation. It can only be satisfactorily explained by arrest of development at a very early stage before the projection of a limb-bud beyond the surface of the germ. The scar and dimple on the right shoulder imply that here a limb-bud had begun to grow out, but was blighted in its very inception. The defects in the right lower limb signify a still later disturbance of development. Differentiation of tissues and skeletal segmentation took place, but the growth of the limb, and in particular of the thigh, was stunted, and articulations were never perfected.

The equino-varus may be regarded as a continuance, in a somewhat exaggerated degree, of the normal condition of the foot at birth, which, as Dr. Little has pointed out, has the sole turned slightly inwards. The persistence and increase of this infantile condition may here be due to the foot never having been brought into use for progression in the erect posture.

The development of the shoulder-blades and clavicles,

the peripheral segments of the limbs being absent, is normal in *Amphisbænidae* (Blindworms), which have a well-developed shoulder-girdle, but do not possess external limbs.*

* *A case of congenital absence of both arms*, was read February 14th, 1873, at the Clinical Society, by Mr. Thomas Smith.—Thomas C—, æt. 4, a robust, healthy boy. No bands or adhesions found when born, and no vestiges of limbs within the membranes; upper limbs entirely absent. A small dimple on each side upon the prominence of the shoulder, over the position of the glenoid cavity; no projecting stump of an arm and no trace of humerus. The clavicles well developed and articulating normally with the acromion on either side; glenoid cavities empty; free movement of shoulders in every direction; can grasp objects between his cheek and his acromion process; prehensile power of his toes well developed; can pick up a small coin easily.

DESCRIPTION OF PLATE VI.

Case of Complete absence of both the Upper Limbs, &c. (J. W.
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