HÉMI-HYPERTROPHIE ALTERNE.

BY

A. DINGWALL FORDYCE, M.D., F.R.C.P.

Physician to the Royal Liverpool Children's Hospital.

It is not uncommon to meet from time to time with cases exhibiting hypertrophy or giantism of some part of the body, of greater or less extent. It may be a part or the whole of a limb, one side of the body, or one half of the face.

The following case is an example of such partial macrosomia.

M. I., female, aged 13 years (Dr. Hartley Martin). Reported "healthy child, but gets fluid in left knee about once a year." When aged 5 years she fell at school and injured her left leg. It was noticed first at this time that the left leg was longer than the right. Since this fall she has had occasionally fluid in left knee.

Fig. 1.—Case of Partial Macrosomia, aged 13 years.

The left leg is obviously bigger and longer than the right (Fig. 1):—

Length of leg : left, 84 cm., right, 80 cm.

Girth of thigh : left, 33·5 cm., right, 31·7 cm.

The reflexes on the two sides are equal.

The synovial membrane of the left knee is greatly thickened. The whole of the left leg shows many superficial dilated vessels, and the soft tissues appear varicose and unhealthy.

X-ray examination proves the existence of true hypertrophy of the left lower limb and healthy bones. Although no abnormality was noted until the age of 5 years, and the condition may consequently be of post-natal origin, it seems more probable that it is congenital, with superadded trauma to the knee arising from instability due to asymmetry.

The above is a case of asymmetry due to true macrosomia, but somewhat similar effects may be produced as a result of atrophy or true microsomia :—

Fig. 2.—Case of True Microsomia, aged 5 years.

J. D., female, aged 5 years (Dr. McFarland). Healthy child. Mother reports that at time of birth it was noticed that the child's left arm and left leg were limp, so much so that the doctor feared they were injured; and that she has always been smaller on the left side than the right. She began to walk and talk at about the age of 15 months, and is apparently mentally normal.

On examination the whole of the left side is at once seen to be smaller than the right (Fig. 2), and X-ray examination proves that this difference affects also the bones. Ossification is equally advanced on the two sides.

The reflexes are normal and appear to be equal on the two sides. The superficial tissues of both sides are healthy, but the mother states that the left leg is often colder than the right.

The measurements of this child are given in Table 1.

TABLE 1.
MEASUREMENTS IN CASE OF MICROSOMIA.

Measurements.							Right.	Left.
Ears: greatest breadth	...	...	...	...	...	...	6.2 cm.	5.2 cm.
Arms: length	...	...	...	...	...	...	46 "	43 "
girth of forearm	...	...	...	...	...	...	15.5 "	14.8 "
Legs: length	...	...	...	...	...	...	56.5 "	54.5 "
girth of thigh	...	...	...	...	...	...	31.8 "	27.4 "
girth of calf	...	...	...	...	...	...	24.4 "	22.2 "
Thorax: hemi-circumferences	...	...	...	...	...	...	28.5 "	27.5 "
Abdomen: hemi-circumferences	...	...	...	...	...	...	28.8 "	26.2 "

It is uncommon to meet with a case of complete unilateral hypertrophy, and a case of *hemi-hypertrophie alterne* or giantism of one half of the trunk and corresponding limbs, with hypertrophy of the face on the opposite side, is very rare.

As the result of exhaustive search into the literature Gesell¹ found that up till April, 1927, 53 cases in all of true unilateral hemi-hypertrophy had been reported. (Of cases recorded since that date may be noted those by McFarland² and Ougrelidze³).

In a previous communication Gesell⁴ gives a table "of thirty cases of partial and crossed hypertrophies" and states in allusion to it "Table 4 is by no means complete, for such partial or restricted hypertrophies are much more common than true total hemi-hypertrophy."

The table includes three cases which would appear to come under the designation crossed hemi-hypertrophy (the cases of Bankhart, Jacobson and Lewen), but in none was the condition that described by André Thomas⁵ as "*hemi-hypertrophie alterne*." Thomas wrote: "*L'hypertrophie faciale peut exister seule, ou bien les membres du même côté sont également hypertrophiés; dans d'autres observations plus rares (Lewen) l'hemi-hypertrophie est croisée. La face et membre supérieur sont hypertrophiés d'un côté, le membre inférieur du côté opposé, il est sans doute exceptionnel que la face soit prise d'un côté, les membres et le tissu du côté opposé; dans leur travail, Sabrazés et Cabannes n'en rapportent aucun cas démonstratif; il s'agit alors en quelque sorte d'une hémihypertrophie alterne.*" In the case described by him there was hypertrophy of the face and tongue on the right side and of the limbs and trunk on the left side.

A case which is difficult to place, and was not quoted by Gesell, was published by Chodak Gregory⁶. In this case, a female aged 2½ years, showed hypertrophy of the left leg, abdomen, thorax, arm and face, with greater semi-circumference of the skull on the right side.

Noronha⁷ described the case of a male Mahomedan, aged about 18, whose face was bigger on the right side, and the trunk and limbs larger on the left, and Slaughter and Eberhardt⁸ reported the case of an American seaman aged 21, who exhibited "an asymmetrical hemi-hypertrophy involving the right side of the face, the right upper extremity and the left lower extremity."

Of these cases, only that of Noronha is similar to the condition coming under the designation "*hemi-hypertrophie alterne*" as described by Thomas and quoted by Ballantyne.⁹

The term hypertrophy is here used in the sense of a true macrosomia—an abnormal largeness of all the tissues of the part involved. There is thus excluded all forms of false hypertrophy, *i.e.*, hypertrophy affecting the soft tissues but not the bones, such as Milroy's disease (congenital or hereditary oedema) and elephantiasis.¹⁰

Ballantyne⁹ defines macrosomia as a monstrous largeness of all the parts of the individual which may have its origin before or after birth, and he prefers the terms 'unilateral macrosomia' to that of 'hemi-hypertrophy.' Acquired hypertrophy, hypertrophy of post-natal origin, is not uncommon of the false type, *i.e.*, where the bone is not affected. It is rarely, even when localized to a comparatively small area, of the true type. (The case described by Simpson¹¹ was that of a true hypertrophy, and may have been post-natal in origin.) The ætiology of true congenital macrosomia is still obscure. Ballantyne stated :

"We may frankly admit that we do not know the causes of unilateral macrosomia or their mode of action; indeed, we do not know those of general macrosomia, and we can hardly, therefore, expect to be more successful in our attempts to arrive at these of the partial variety of giant growth. . . . From the standpoint of ante-natal pathology we have only to do with the congenital cases; and among them there are some which may be truly embryonic in origin while others are apparently foetal. It is often exceedingly difficult to separate the giant growths which are really due to an original anomaly of growth from those which arise in the foetal period of ante-natal life and are caused by such diseases as congenital elephantiasis and angiomas. . . . The causes which are usually alleged in connection with all teratological cases are also advanced to explain unilateral macrosomia of the head and face; and the theory that in some way the overgrowth is due to perverted action of the nervous system (foetal meningitis) would appear to be the most probable. On the other hand, there is much to suggest that the macrosomia originates before the foetal period of life, and therefore before foetal diseases can be invoked as causative agencies. Our knowledge of the teratogenesis of this, as of all forms of macrosomia and microsomia, is, after all, close neighbour to nothing."

It has been noted by several observers, among them Lockhart Mummery,¹² that true congenital hypertrophy of one half of the body and face has been associated with a greater size of the cranium on the opposite side. A photograph of Mummery's patient appears in Purves Stewart's book on the Diagnosis of Nervous Diseases, and in allusion to it the author states that the condition was "probably of cerebral origin . . . the right side of the cranium and presumably also the right side of the brain were larger than the left," while the hypertrophy of body and face was on the left side. It is obvious that such a suggestion is by no means an exposition of the fundamental ætiology, and in recent years the most interesting and intriguing opinion is that of Gesell (*loc. cit.*). He considers that hemi-hypertrophy, while being a "morphogenetic anomaly dating back to an early embryonic stage," is to be regarded as a "minimal form of twinning." "It is a unilateral enlargement of one-half of the soma, a hemi-macrosomia. As such we may interpret the condition to be an atypical or a paradoxical form of twinning, a hybrid variant of the same process which may produce a double monster or a completely symmetrical individual. The biologic paradox consists in this, that the hemi-hypertrophy is neither double monstrosity nor bilateral duplicity; it is half of each, as though the individual remained two conjoined hemi-creatures, each with a discrete though half realized genetic destiny."

As Ballantyne says, "In all speculations about the ætiology of anomalies of growth, it has constantly to be borne in mind that a theory to be satisfactory must account for the facts, and for all the facts." Cases of *hémihypertrophie alterne* do not render theorizing any simpler.

Before reporting the following case which appears to be one of this type of macrosomia, two recent publications may be alluded to.

Ilse Graetz¹³ describes a case from the Children's Clinic of the University of Kiel of "sogennanter 'totaler halbseitiger Körperhypertrophie.'" In this case (a girl of 1½ years) the whole of the left side was more developed than the right, but whereas the hypertrophy of the leg was a true hypertrophy, that of the arm with the exception of two fingers was proved by X-ray examination to be a hypertrophy solely of the soft tissues.

In discussing the case he writes: "Es handelt sich bei der sog. Halbseitigen Hypertrophie gar nicht um eine "einfache Hypertrophie" der einen Körperhälfte, gleich als ob das Individuum aus zwei zwar zueinander nicht passenden, aber in sich 'normalen' Hälften bestände, von denen es fraglich sein könnte, welche von beiden Hälften die (für das betreffende Individuum) 'normale,' welche die 'pathologische' sei, sondern die sog. halbseitige Hypertrophie trägt in allen genau untersuchten Fällen stets den *Charakter einer Missbildung*." Later he states: "In allen genau untersuchten Fällen zeigten die hypertrophierten Organe einen—mehr oder minder—pathologischen Befund," and the last words in his concluding summary are "Bei allen genau durchuntersuchten Fällen von partiellem wie halbseitigem Riesenwuchs handelt es sich nie um eine 'reine Hypertrophie,' sondern stets um dystrophische Veränderungen der hypertrophierten Organe."

This case of Graetz is interesting in its apparent mixture of true and false hypertrophies, but its study seems to have carried the author off his feet and into the use of those dangerous words "always" and "never." Further, he appears to consider that no English case has been "genau durchuntersucht," as no English author is quoted in his extensive bibliography. This seems a pity, as an evening with Ballantyne may be both illuminating and chastening.

The other report is that by Wakefield¹⁴. He reports a case of congenital false hemi-hypertrophy of the right side in a girl of 6 years. When discussing ætiology he remarks: "Practically all the exponents of the many theories on this anomaly, except one (Gesell) have been content to advance their theory and then silently repent. In the future when fashionable medical thought turns to something more obscure than embryology we are sure to have more interesting theories on this anomaly."

Hémi-hypertrophie Alterne.

CASE REPORT :

J. F., female, born February, 1924; examined 16th August, 1928. Height, 95 cm., weight 36½ lb. (Dr. Foster, Birkenhead). Son of maternal aunt has syndactyly. No history of twins in any part of the family.

Parents healthy. Mother reports fall at fifth month of pregnancy. Two other children, both girls, aged respectively 8 years and $2\frac{1}{2}$ years: both well formed and healthy. No miscarriages.

Noticed at birth there was 'a water bladder like a balloon on the navel.' The baby appeared quite healthy, but it was evident at birth that the right side of the body and the right arm and leg were bigger than the left; while the left side of the face and left side of the tongue were

Fig. 3.—Case of *Hémi-hypertrophie alterne*, aged 4 years.

bigger than the right. The differences were as marked at birth as at 4 years old. She has been a fairly healthy child and had no serious illness. She did not walk till she was 3 years old and talks very little even now. There is an indefinite history of fits in babyhood, but delay in walking and talking is largely explicable by disparity in size of legs and of two sides of tongue. She is, however, definitely mentally retarded, although her mother considers her quite sensible.

No vascular or trophic changes are detectable; temperature, sensibility and perspiration are similar on the two sides. The reflexes are equal on the two sides.

Measurements.

Head circumference, 46 cm., right half, 23 cm., left half, 23 cm.

Vault of skull to posterior angle of jaw : left, 17 cm., right 15 cm.

Ear to mid-line of lip : left, 11 cm., right, 10.5 cm.

Teeth : 5 above and below on either side.

Left half of tongue much larger than right, and left cheek, ear and face generally noticeably larger than right.

Arm : total length (acromion to tip of mid-finger), right, 47 cm., left 42 cm.

Upper arm : length, right, 24 cm., left, 20 cm.

girth, right, 17 cm., left, 16 cm.

Lower arm : length, right, 16 cm., left, 14 cm.

Chest : hemi-circumference, right, 32 cm., left, 29 cm.

Abdomen : hemi-circumference, right, 32 cm., left, 27 cm.

Leg : total length, right, 51 cm., left, 45 cm.

Thigh : length, right, 27 cm., left, 24 cm.

girth, right, 31 cm., left, 21 cm.

Lower leg : length, right, 24 cm., left, 21 cm.

Foot : length, right, 17 cm., left, 15 cm.

Right labium markedly larger than left.

No abnormality or peculiarity distinguishable as regards internal organs.

X-ray examination shows increased size of the bones of the right side with advance in ossification (see Fig. 4 and 5).

DISCUSSION.

In many of the reported cases of macrosomia and hemi-hypertrophy, nævi, cutaneous congestion, or other superficial deviations from the normal have been noted. These are frequently on the hypertrophied side of the body or on both sides, but a particularly interesting case with reverse localization is reported by Babonneix.¹⁴

In some cases dentition is noted as being more advanced on the hypertrophied side ; and the same is sometimes true of ossification.

While the general health is usually good, many of the cases are reported as being feeble-minded. In the case reported by Thomas, however, first seen when aged 4 months and later when 27 months, it is noted that the mentality was normal and the physical differences had, during the interval between observations, become rather less.

What does the future hold in store for J. F., and on what lines should she be treated ?

She has been under observation nearly 12 months, and although during this period her physical disproportion has not increased, it has not to any appreciable extent diminished.

We know that in some cases of this kind, with the passage of time, an approach towards normal symmetry occurs, and from the physical aspect there is consequently no call for pessimism. With the assistance of the orthopædist she should grow up if physically incommoded, yet not seriously either incommoded or disfigured. The seat of gravity in prognosis lies deeper. She is feeble minded, and she will certainly " be permanently incapable by reason of such defectiveness of receiving proper benefit from the instruction in ordinary schools " (Mental Deficiency Act, 1927).

Fig. 4.—*Hémi-hypertrophie alterne*: skiagram showing increased size of bones and advanced ossification on right side.

Fig. 5.—*Hémi-hypertrophie alterne*: skiagram showing increased size of bones and advanced ossification on right side.

Suitable education and training is here a matter of medical direction, and such direction must be away from the Ordinary Elementary School and the Special School for Physically Defective Children, and towards the Special School for Mentally Defective Children. Orthopædic ailments or defects not infrequently are so obtrusive from the purely orthopædic aspect, that the basal lesion or its non-orthopædic results or concomitants pale in comparison.

It is but a step from an orthopædic clinic to a P. D. School. But orthopædic conditions resulting from intracranial trouble, be it congenital or post-natal, developmental or acquired, in most instances are but one form of expression of a double defect, and the obvious orthopædic defect is of comparatively minor importance. The other form is feeble-mindedness, and the clamant call is for appropriate education.

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