subclinical viral hepatitis. Consequently there should be little risk in otherwise healthy long-term carriers of HBA visiting barbers or dentists or working as nurses, physicians, and so on. Earlier opinions on this problem (Chalmers and Alter, 1971) can probably be modified provided that the results of this study are confirmed.

References

- Baggenstoss, A. H. (1957). *Journal of the American Medical Association*, 165, 1099.
- Bancroft, W. H., Mundon, F. K., and Russell, P. K. (1972). *Journal of Immunology*, 109, 842.
- Bianchi, L., et al. (1971). *Lancet*, 1, 333.
- Chalmers, T. C., and Alter, H. J. (1971). *New England Journal of Medicine*, 285, 613.
- Cherubin, C. E. (1971). *Lancet*, 1, 627.
- Groote, J. de, et al. (1968). *Lancet*, 2, 626.
- Holland, P. V., Purcell, R. H., Smith, H., and Alter, H. J. (1972). Hepatitis Scientific Memoranda (Memo H-283), NIH, USA.
- Iwarson, S., and Hermodsson, S. (1971). *Scandinavian Journal of Infectious Diseases*, 3, 93.
- Iwarson, S., Lindholm, A., Lundin, P., and Hermodsson, S. (1972). *Vox Sanguinis*, 22, 501.
- Kim, C. Y., and Tilles, J. G. (1971). *Journal of Infectious Diseases*, 123, 618.
- Krugman, S., and Giles, J. P. (1970). *Journal of the American Medical Association*, 212, 1019.
- Le Bouvier (1971). *Journal of Infectious Diseases*, 123, 671.
- Lundvall, O., and Iwarson, S. (1970). *Acta Medica Scandinavica*, 187, 225.
- Levene, C., and Blumberg, B. S. (1969). *Nature*, 221, 195.
- Magnius, L.-O., and Espmark, A. (1972). *Journal of Immunology*, 109, 1017.
- Magnius, L.-O., Espmark, A., and Ringertz, O. (1972a). *Acta Pathologica et Microbiologica Scandinavica*, 80, 340.
- Magnius, L.-O., Berg, R., Bjorvatn, B., Espmark, J. A., and Svedmyr, A. (1972b). In preparation.
- Mosley, J. W., Edwards, W. M., Neikans, J. E., and Reidecher, A. G. (1972). *American Journal of Epidemiology*, 95, 529.
- Schmidt, N. J., Roberto, R. R., and Lennette, E. H. (1972). *Infection and Immunity*, 6, 1.

MEDICAL MEMORANDA

Congenital Hemihypertrophy with Aortic, Skeletal, and Ocular Abnormalities

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Congenital hemihypertrophy is a rare malformation in which one half of the body is more developed than the other.

Case Report

A woman aged 24 was found on routine premarital examination to have a cardiac murmur and anomalies of the limb extremities. There was no history of previous illness although she walked with a slight limp.

She was admitted to hospital for investigation. On examination there was striking asymmetry of the body (fig. 1), particularly the upper part. The right upper limb was 4 cm longer than the left and was thicker. The difference was less noticeable in the lower limbs, the right leg being 3 cm longer than the left. The face showed a mild asymmetry; the right eye was higher than the left, the right ear larger, the right part of the lip thicker, and the chin more prominent on the right side. The teeth and tongue were symmetrical. The left iris was more deeply coloured than the right. Several fingers of the left hand were shortened. There was deformity of the big toes together with syndactyly of the second and third toes on each side. The right breast was larger than the left. Radiographs of the skeleton showed that on the right side each bone was thicker and longer than its counterpart on the left. In addition, the right femoral head was incompletely covered by the acetabulum. Films of the hands (fig. 2) showed, on the left side, shortening of the first metacarpal and first phalanx of the thumb and clinodactyly of the first and fifth fingers. The cubital styloid was absent. Radiographs of the feet (fig. 3) showed that on the right side there was clinodactyly of the second phalanx of the great toe, absence of the second phalanx of the fifth toe, and shortening of the second phalanx of the other toes. On the left foot there was shortening of the metatarsal bones and of the first phalanges.

Blood pressure was 150/70 mm Hg in the right arm and 110/70 mm Hg in the left. There was a rough systolic murmur situated high up in the aortic area below the clavicle. It was of medium intensity and radiated into the neck. The second sound

FIG. 1—Patient, showing right-sided hemihypertrophy.

FIG. 2—Radiographs of hands.

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was accentuated in the aortic area. The electrocardiogram was normal. The phonocardiogram showed an ejection systolic murmur in the aortic area and confirmed the loud aortic second sound. The chest film showed a normal heart and a normal vascular pattern. The aortic knuckle was small, suggesting that the aorta was on the right side. Barium-swallow examination confirmed this hypothesis, showing a posterior and right oesophageal notch.

A clinical diagnosis of supra-valvular aortic stenosis was made. Left heart catheterization confirmed the right-sided aortic arch but showed no evidence of aortic stenosis. The endocavitary phonocardiogram showed a systolic murmur localized to the origin of the left subclavian artery, which was implanted low on the

FIG. 3—Radiographs of feet.

FIG. 4—Angiogram showing right-sided aorta and anomalous origin of left subclavian artery.

descending aorta. Angiography (Fig. 4) showed that the aorta was right-sided and that the left subclavian artery arose from an aortic diverticulum which was located below the usual level of the origin of this artery. This abnormality has been described by Gross and Ware (1946) and fits into Type B2 C of their classification. The diverticulum is a remnant of the distal part of the left aortic arch. In association with this abnormal origin the first part of the left subclavian artery was tortuous and irregular, suggesting a possible site of origin for the murmur. It was also noticed that the left vertebral artery was not opacified.

Ocular abnormalities were also present, consisting of ectropion of the ectodermal leaflet of the left iris with a right iridic cyst and abnormal pigmentation.

Comment

The diagnosis of congenital hemihypertrophy is usually made in infancy or childhood. The present case is notable not only

for the late diagnosis but also, particularly, for the associated abnormalities of the aortic arch, with a right-sided aorta and low implantation of the left subclavian artery, which arose from an aortic diverticulum. Another interesting point was the presence of anomalies of the extremities, brachydactyly, and syndactyly.

Hemihypertrophy is slightly more frequent on the right side than on the left (in the proportion 53:47 according to Ringrose *et al.* (1965)). Associated abnormalities are frequent (Carron, 1951; Gerloczy *et al.*, 1965; Ringrose *et al.*, 1965) but vascular abnormalities are usually superficial; three cases of congenital cardiomyopathy have been recorded by Ringrose *et al.*, but no abnormality of the aortic arch has been noted previously.

Diagnosis is generally easy, although in mild cases, as in the one reported here, measurements of each part of the body may be necessary. In differential diagnosis the congenital hemiatrophies, which are rare and usually associated with disease of the nervous system, must be excluded, as must localized acquired hemihypertrophy, which may result from vascular abnormalities. The most difficult differential diagnosis is from phacomatosis (neuroectodermal dysplasia), which consists of developmental anomalies of the brain, skin, retinae, bones, and viscera and is hereditary and familial.

PATHOGENESIS

There are three principal hypotheses:

(1) According to Korretas (cited by Charocopoulos and Cocolis (1955)) the hypothalamus is a morphogenetic centre, and any teratogenic influence on the hypothalamus early in embryonic life may induce hemihypertrophy.

(2) Gesell (1921) believed that hemihypertrophy is an abnormality of early development occurring at the beginning of the first segmentation of the primordial follicle.

(3) A third hypothesis postulates a chromosomal abnormality. Some familial cases have certainly been described. Moreover, two cases of hemihypertrophy have been reported (Ellis *et al.*, 1963; Ferrier *et al.*, 1964) which were associated with a diploid-triploid mosaic, and in each case the diploid and triploid cells were found in the skin but the blood contained only diploid cells. Hook and Yunis (1965) reported a case of congenital hemihypertrophy associated with mosaicism and trisomy 18; normal and abnormal cells were found in both halves of the body in equal proportions.

PROGNOSIS AND TREATMENT

Asymmetry between the two halves of the body is fixed at the end of bone growth. By this stage only palliative treatment is available. The aim is to compensate for the shortening of one lower limb in order to prevent static spinal disorders. This may be done with a built-up shoe or a heel piece, but if one limb is markedly longer than the other a corrective partial osteotomy may be necessary.

References

- Carron, R. (1951). *Pédiatrie*, **6**, 969.
- Charocopoulos, S., and Cocolis, J. (1955). *Pédiatrie*, **10**, 709.
- Ellis, J. R., Marshall, R., Normand, I. C., and Penrose, L. S. (1963). *Nature*, **198**, 411.
- Ferrier, D. (1964). *Lancet*, **1**, 80.
- Gerloczy, F. *et al.* (1965). *Acta Paediatrica Academiae Scientiarum Hungaricae*, **6**, 423.
- Gesell, A. (1921). *Archives of Neurology and Psychiatry*, **6**, 400.
- Gross, R. E., and Ware, P. F. (1946). *Surgery, Gynecology and Obstetrics*, **83**, 435.
- Hook, F. B., and Yunis, J. J. (1965). *American Journal of Diseases of Children*, **110**, 551.
- Ringrose, R. E., Jabbour, J. T., and Keele, D. (1965). *Pediatrics*, **36**, 434.