process such as disseminated sclerosis. Nor was there any evidence to support a diagnosis of toxic or nutritional encephalopathy.

That the condition was polioencephalitis, which can at times be extremely difficult to differentiate from encephalitis lethargica (Brewis and Neubauer, 1948) and may present with almost any combination of brain-stem and cranial nerve signs (McAlpine, 1947), is unlikely in view of the absence of evidence of involvement of anterior horn cells and the negative virus and serum antibody studies in Cases 2 and 3. The prolonged initial illness, the C.S.F. findings, and the nature of the sequelae are also much against this diagnosis. Furthermore, the three cases reported here occurred at a time when poliomyelitis was not locally epidemic, and there were no other cases of encephalitis of any type notified in the military or civilian population of Northern Germany at this time.

An illness similar to encephalitis lethargica is only very rarely produced by the many known types of virus encephalitis, and ophthalmoplegia and Parkinsonism are conspicuous by their absence, except in von Economo's disease (Schlesinger, 1952). Moreover, many of the other types are summer infections, whereas the seasonal incidence of encephalitis lethargica is strikingly different, the majority of cases occurring in the first three or four months of the year (Ministry of Health Report, 1922; von Economo, 1931), as did the cases reported here. Virus studies in Cases 2 and 3 were negative for three neurotropic viruses, in addition to poliomyelitis. Further work is required to determine the aetiology of clinical encephalitis lethargica and to show whether it can be produced by more than one organism. At the present time, while the diagnosis remains a clinical one, it must be applied to these and similar cases.

Summary

Three cases which occurred recently have been diagnosed as encephalitis lethargica. They add further evidence that this disease still occurs and that post-encephalitic Parkinsonism may be due to recent infection.

The diagnosis is discussed. If the characteristic clinical picture and sequelae of this disease are observed, there can be no justification for rejecting the diagnosis on the doctrinaire ground that encephalitis lethargica is a disease of the past. The diagnosis must be based on purely clinical criteria.

We are grateful to Dr. P. Mallam for permission to publish Case 1, and to the Director-General, Army Medical Services, for permission to publish Cases 2 and 3. We wish to thank Dr. W. Ritchie Russell and Dr. C. W. M. Whitty for their advice and assistance, and Dr. A. D. Macrae for the virus studies in Cases 2 and 3.

REFERENCES

- Barrett, A. M., Gairdner, D., and McFarlan, A. M. (1952). *British Medical Journal*, 1, 1317.
 Bickerstaff, E. R., and Cloake, P. C. P. (1951). *Ibid.*, 2, 77.
 Brewis, E. G. (1954). *Ibid.*, 1, 1298.
 — and Neubauer, C. (1948). *Ibid.*, 2, 416.
 Buzzard, E. F., and Greenfield, J. G. (1919). *Brain*, 42, 305.
 Ford, F. R. (1946). *Diseases of the Nervous System in Infancy, Childhood and Adolescence*. Thomas, Springfield, Illinois.
 Garland, H. G. (1953). In *Medicine*, vol. 2, edited by H. G. Garland and W. Phillips. Macmillan, London.
 Hall, A. J. (1924). *Epidemic Encephalitis*. Wright, Bristol.
 Hoare, R. D. (1953). *Acta radiol. (Stockh.)*, 40, 96.
 Leigh, A. D. (1946). *British Medical Journal*, 2, 937.
 Logue, V., and Monckton, G. (1954). *Brain*, 77, 252.
 McAlpine, D. (1947). *Proc. roy. Soc. Med.*, 40, 929.
 Ministry of Health (1922). Report on Public Health and Medical Subjects. No. 11: Encephalitis Lethargica. H.M.S.O., London.
 Neal, J. B. (1942). *Encephalitis—a Clinical Study*. Grune and Stratton, New York.
 Schlesinger, R. W. (1952). In *Monographs in Medicine*, Series 1. *Virus Encephalitis*, p. 380, edited by W. Bean. Williams and Wilkins, Baltimore.
 Taylor, L. M., Smith, H. V., and Hunter, G. (1954). *Lancet*, 1, 700.
 Teilmann, K. (1953). *A.M.A. Arch. Neurol. Psychiat.*, 69, 208.
 von Economo, C. (1931). *Encephalitis Lethargica: its Sequelae and Treatment*. Oxford Univ. Press, London.
 Wartenberg, R. (1945). *The Examination of Reflexes*. Year Book Publishers, Chicago.
 Wilson, S. A. Kinnier (1940). *Neurology*, edited A. N. Bruce. Arnold, London.

CONGENITAL ABNORMALITIES OF THE UTERUS AND PREGNANCY

BY

JOHN A. HOLMES, M.B., M.A.O., M.R.C.O.G.

Late Registrar, St. James' Hospital, London, S.W.12

Pregnancy associated with congenital uterine malformation is not uncommon. However, it has been given little attention in the standard textbooks of obstetrics. Isolated cases have been reported, but few papers on the subject have been published. In the maternity department of Farnborough Hospital, Kent, I had the opportunity of observing six cases. Three further cases were observed in the Obstetric Department of St. James' Hospital, London. This paper is the outcome of interest aroused by these cases. The cases are reported briefly, and some aspects of the association of congenital uterine malformations and pregnancy are discussed.

The earliest reference to the subject is a case reported by Mauriceau in 1675. Isolated case reports followed this, and Kussmaul published a paper on the condition in 1859. Miller (1922) summarized the literature up to that time and Smith (1931) reported 35 cases. In the last decade papers have been presented by Jarcho (1946), Fenton and Singh (1952), and Baker *et al.* (1953). In the British literature, Way (1945) and Hunter (1950) published papers describing the condition.

Incidence.—In the cases reported from Farnborough Hospital, the six latest pregnancies occurred among 5,000 deliveries, giving an incidence of 0.12%. The three latest pregnancies in the cases seen in St. James' Hospital were among 3,500 deliveries, giving an incidence of 0.08%. From the literature it would appear that the incidence of the association of this abnormality with pregnancy varies greatly. Smith (1931) gives an incidence of 0.003%, Way (1945) reports an incidence of 0.014%, while Baker *et al.* (1953) give 0.03%. It would seem that 1 in 1,000 would be a reasonably correct figure.

Development.—The Müllerian ducts appear around six weeks as two longitudinal folds on the dorsal aspect of the coelomic cavity. They are situated on the outer aspect of the intermediate cell mass, lateral to the genital ridge and Wolffian system. The ducts turn inwards, cross internal to the Wolffian ducts, and lie side by side in the midline. The lower portions fuse at about 14 weeks and they push into the urogenital sinus to form the Fallopian tubes and the uterovaginal canal. Anomalies arise because of: (1) failure of fusion (types A, B, and C); (2) persistence or partial persistence of the septum between them (types D, E, and F); (3) failure of one side to develop (type G).

Classification.—The classification I have followed is shown in Fig. 1. It is adapted from Jarcho (1946). The classification is a simple one and embraces all the common types. As is shown later, the proper classification of a congenital abnormality is of great importance where pregnancy is concerned.

Other Congenital Abnormalities.—Where abnormality of the uterus exists it is probable that other abnormalities will be found, chiefly involving the structures developed from the intermediate cell mass. Thus there is often associated congenital abnormality of the urinary tract, and sometimes congenital abnormalities in other parts of the body. Associated urinary tract anomaly is so

frequent that investigation of the patient is incomplete without an intravenous pyelogram. This often reveals absence of a kidney or ectopic kidney. An intravenous pyelogram was done in six cases, and a double ureter was

anaesthesia and curettage performed. Baker *et al.* (1953) advise utero-salpingography in such cases to exclude the presence of a congenital uterine abnormality.

Ante-partum Complications

Miller (1922) drew attention to the higher incidence of threatened and inevitable abortion in cases where pregnancy is associated with congenital uterine anomalies. In the small number of cases reported, the abortion rate is noticeably high in types C, D, and E, and this corresponds with the findings of previous writers. Thus, in Case 5, reported below (Type C) (Fig. 3), four abortions occurred before the patient had a viable pregnancy, while in Case 8 (Fig. 4) two abortions preceded a viable pregnancy. This case has been classified as type F, but the presence of a small septum could not be ruled out. Way (1945), discussing types C, D, and E, reported 12 abortions in 33 pregnancies. The probable explanation for the increased

FIG. 1.—Classification of types of congenital uterine abnormalities. A, Uterus pseudodidelphys. B, Uterus duplex bicornis bicollis; vagina simplex. C, Uterus bicornis unicollis; vagina simplex. D, Uterus septus. E, Uterus subseptus. F, Uterus arcuatus. G, Uterus unicollis. (After Jarcho, 1946.)

found in one (Case 3) (see Fig. 2). Keevil (1943) and Burton-Browne (1948) have published cases demonstrating the association of genital and urinary tract anomalies.

Diagnosis

The first essential for the diagnosis of a congenital uterine abnormality in pregnancy is that the accoucheur should bear the condition in mind. The diagnosis is most easily made in early pregnancy. On routine vaginal examination the presence of a vaginal septum or two cervices may be detected, as in Cases 1, 2, 6, 7, and 9. On bimanual examination the accessory uterine horn may be palpated. The differential diagnosis will be from extra-uterine pregnancy, extrauterine tumour, or pregnancy associated with fibroids. Occasionally the diagnosis will be known before the pregnancy began, as a result of utero-salpingography (Case 8) or laparotomy (Case 4). In the later months the persistence of a transverse lie, particularly in a primigravida, should arouse suspicion of the condition. This is stressed by Way (1945), particularly with abnormalities of types C, E, and F. In cases with a history of recurrent abortion or recurrent premature labour the condition should be considered and examination under

FIG. 3.—Hystero-gram from Case 5, showing type C abnormality.

abortion rate is that implantation may occur on the septum, and this is an unfavourable nidation site. In types A and B the abortion rate is not increased. Baker *et al.* (1953) give the incidence in these types as only 8.2%, while there were no abortions in our cases. Often in types A and B bleeding may occur in early pregnancy; it was noticed in these types in three out of the six most recent pregnancies in this series. This bleeding is probably due to shedding of the endometrium in the non-pregnant horn. Occasionally a uterine cast from the non-pregnant side is shed.

Malpresentations

Malpresentations are common. The more complete the malformation the less likely is malpresentation to occur (Kerr and Moir, 1949). In the more complete types—that is, A and B—the foetus must assume a longitudinal lie, so that the presentation is vertex or breech, and transverse lie does not tend to occur. Breech presentation is therefore more common in these types. It occurred twice in eight pregnancies in our cases of types A and B. In Fenton and Singh's (1952) cases the incidence of breech presentation was 10%. In types C and E, however, transverse lie is also common. Way reports its occurrence 12 times in 32 pregnancies, while breech presentation was recorded four times. In three of our cases breech presentation recurred in a second pregnancy. Twins are more common in congenital anomalies of the uterus (Berkeley *et al.*, 1938). Cases have been reported where each foetus occupied a separate uterine horn (Williams and Cummings, 1953; Moore, 1936).

FIG. 2.—Hystero-gram from Case 3, showing type A abnormality.

Premature labour is more common in all types of congenital abnormality. Of the 12 pregnancies beyond the 28th week, reported below, premature labour occurred in 5, and in 4 the infants were under 5½ lb. (2.5 kg.). Fenton and Singh (1952) maintain that premature rupture of the membranes and premature labour are twice as common as in normal pregnancy. They regard this as being due to the irregular uterine contour and to the increased incidence of malpresentations. A uterine cast may be shed from a non-pregnant horn at any time during the antenatal period. Such a case has been reported by Corbet (1946).

FIG. 4.—Hystero-gram from Case 8, showing type E abnormality.

Complications in Labour

Premature labour has already been mentioned. Persistent transverse lie will cause difficulty in types C, D, and E. This complication was not met with in the present cases. Way stresses it and mentions the additional hazard of prolapse of the cord. Breech delivery occurs more frequently in all types. In the second stage of labour delay may be caused by a vaginal septum which has become thick and oedematous during labour. In three out of the four cases of type A reported, it was necessary to incise the septum as it was obstructing delivery. This was done under local analgesia with 1% lignocaine hydrochloride, and this procedure is recommended. It is not painful, nor is it attended by much bleeding. Fenton and Singh (1952) also report that where the septum was present it was necessary to incise it during delivery in almost all cases.

All writers draw attention to the risk of the non-gravid horn becoming impacted and obstructing labour. Dougal (1921) reported such a case. Fenton and Singh (1952) did not encounter this difficulty in 146 collected cases, and it was not met with in the present cases. It is probably a very rare complication. In all types of congenital uterine abnormality there is increased risk of retained placenta and post-partum haemorrhage. There were 10 vaginal deliveries in the cases reported. Primary post-partum haemorrhage occurred in four and secondary post-partum haemorrhage in one. Manual removal of the placenta was carried out three times. The increased incidence of post-partum haemorrhage and manual removal of the placenta was present, irrespective of the type of congenital abnormality. Hunter (1950) reported a manual removal rate of 40%. During the puerperium a cast of a non-pregnant horn may be shed. It was noticed in one of our cases.

Maternal and Foetal Hazards

Apart from rupture of a pregnancy in a rudimentary horn, the presence of congenital uterine abnormalities does not appear to increase the maternal mortality. Two deaths are recorded by Baker *et al.* (1953), but these were due to

rupture of a rudimentary uterine horn. This condition presents rather the features of an extrauterine gestation, and is not considered in this paper. A morbid puerperium was recorded in four cases in the series here recorded. Most authors give the morbidity rate as 20%. This high figure is undoubtedly due to the increased number of manipulations required during labour in these cases.

The foetal mortality is again related to the type of abnormality. I had one stillbirth in 10 deliveries in types A and B. Baker *et al.* (1953) give a 4.3% foetal mortality in this group, while 5.5% is the figure quoted by Fenton and Singh (1952). In types C, E, and F, however, Way (1945) gives a 34% foetal mortality, and Baker *et al.* (1953) give the figure as 23.9%. In the present cases there was one stillbirth in three pregnancies. It would thus seem that, while the prognosis in types A and B is good, a pregnancy in types C, E, and F is fraught with danger to the foetus. This is due to the fact that in types C, E, and F shoulder presentation with its attendant risks is more common. Prematurity is a common hazard in both groups, and Fenton and Singh (1952) mention that 25% of the infants are under 5½ lb. (2.5 kg.). There is an increased risk of congenital abnormalities of the foetus, the incidence being around 3%. One case is recorded in this series: a stillborn female infant was found to have, among other deformities, a congenital uterine abnormality of type C.

Caesarean Section

There were three caesarean sections in the cases. Two were performed because of breech presentation in type A abnormality, and one because of four previous miscarriages, in a patient with type C abnormality. In view of the very high foetal mortality in types C, E, and F, mentioned above, it seems reasonable to consider elective caesarean section in this group, particularly if there is persistent oblique or transverse lie. In abnormalities of type A and B the foetal mortality is low, the foetus lies longitudinally, and spontaneous delivery is the rule. In these types an expectant attitude to spontaneous delivery is indicated and there is little place for elective caesarean section. Hoffman (1946) supports elective caesarean section for all types. Baker *et al.* (1953) and others state that it has no place in the management. I hold that the mode of delivery must be influenced by the type of abnormality present, and that there is little place for elective caesarean section in types A and B even when the breech presents.

Case Reports

Case 1.—Aged 21. Type A. Diagnosis: A vaginal septum and two cervices were found on vaginal examination during her first pregnancy. First pregnancy, 1950: This was in the right horn of the uterus and proceeded normally. The breech presented at 32 weeks' gestation and persisted. At 38 weeks the membranes ruptured, and classical caesarean section was carried out because of breech presentation and a double uterus. The infant weighed 6 lb. 11 oz. (3,030 g.). The puerperium was morbid. This was due to a *Staphylococcus aureus* infection of the birth canal. Second pregnancy, 1953: This was in the left horn. Threatened abortion occurred at 10 weeks. After this the pregnancy proceeded normally. Labour occurred at 35 weeks. Spontaneous vertex delivery followed episiotomy and division of the vaginal septum under local analgesia. The third stage was normal. The infant weighed 4 lb. 15 oz. (2,240 g.). The puerperium was not morbid.

Case 2.—Aged 21. Type A. Diagnosis: A septate vagina and two cervices were found at 18 weeks' gestation. Pregnancy, 1954: This was in the left horn and proceeded normally. At 28 weeks a small ante-partum haemorrhage occurred. There was no further complication. Labour began at 41 weeks. Spontaneous vertex delivery followed episiotomy and division of the septum under local analgesia. The infant weighed 7 lb. 1 oz. (3,200 g.). The third stage of labour was normal. The puerperium was morbid because

of a *Bact. coli* infection of the birth canal. A decidual cast of the non-pregnant horn was passed on the fourth day.

Case 3.—Aged 26. Type A. Diagnosis: A double vagina and two cervices were found at booking examination. First pregnancy, 1950: Miscarried at 12 weeks' gestation. Second pregnancy, 1953: This was in the left horn. Threatened abortion occurred at 12 weeks, otherwise the antenatal period was normal. Spontaneous delivery occurred at 40 weeks. The infant weighed 7 lb. 7 oz. (3,370 g.). There was post-partum haemorrhage of 22 oz. (625 ml.). The puerperium was not morbid.

Case 4.—Aged 26. Type C. Diagnosis: Laparotomy was carried out at 8 weeks' gestation, as extrauterine gestation was suspected. First pregnancy, 1949: Spontaneous delivery at 39 weeks of a 5-lb. (2,270-g.) female infant at another hospital. The third stage of labour was normal. Second pregnancy, 1953: Threatened abortion occurred at 6 weeks, otherwise the antenatal period was normal. Spontaneous delivery of a 6 lb. 6 oz. (2,890 g.) female infant occurred at 40 weeks. There was post-partum haemorrhage of 50 oz. (1,420 ml.). Manual removal of the placenta was carried out. The puerperium was normal.

Case 5.—Aged 30. Type C. Diagnosis: This was made during evacuation of an incomplete miscarriage. First pregnancy, 1943: Abortion occurred at 8 weeks' gestation. Second pregnancy, 1947: Abortion occurred at 10 weeks. Third pregnancy, 1949: Abortion occurred at 12 weeks. Evacuation of the uterus was done as the abortion was incomplete. Fourth pregnancy, 1951: Abortion occurred at 22 weeks. Fifth pregnancy, 1953: The antenatal period was normal up to 32 weeks. Threatened premature labour occurred at that time. The patient was delivered by classical caesarean section at 38 weeks, the indications being congenital uterine abnormality and a bad obstetric history. The infant weighed 5 lb. 12 oz. (2,610 g.). The puerperium was normal.

Case 6.—Aged 35. Type A. Diagnosis: A double vagina and two cervices were found on examination at 40 weeks' gestation. First pregnancy, 1955: The patient was first seen at 40 weeks, when she was admitted to hospital with pre-eclamptic toxæmia. The pregnancy was in the left horn. The foetus presented as a breech with extended legs. Classical caesarean section was carried out, the indication being breech presentation associated with congenital uterine abnormality and pre-eclamptic toxæmia. The infant weighed 7 lb. 2 oz. (3,230 g.). The puerperium was morbid because of a *Staph. aureus* infection in the birth canal.

Case 7.—Aged 22. Type A. Diagnosis: A double vagina and two cervices were found on vaginal examination. Pregnancy, 1954: This was in the right horn. The antenatal period was uneventful. The patient went into labour at 40 weeks' gestation. Foetal distress was diagnosed in the second stage. Spontaneous vertex delivery occurred shortly afterwards. The infant was stillborn and weighed 7 lb. (3,175 g.). The cause of death was not apparent. Manual removal of the placenta was carried out because of retained placenta without haemorrhage. The puerperium was normal. Permission was not granted for post-mortem examination of the baby.

Case 8.—Aged 23. Type E. Diagnosis: A type E uterine abnormality was diagnosed at utero-salpingography. First pregnancy, 1950: Abortion occurred at 12 weeks' gestation. Second pregnancy, 1951: Abortion occurred at 12 weeks. Third pregnancy, 1952: The early antenatal period was normal. Breech presentation was diagnosed at 34 weeks. External cephalic version was not successful. Spontaneous breech delivery occurred at 40 weeks. The infant weighed 5 lb. 8 oz. (2,495 g.). Secondary post-partum haemorrhage occurred in the puerperium, necessitating evacuation of the uterus and blood transfusion. Fourth pregnancy, 1953: The early antenatal period was uneventful. Premature labour occurred at 28 weeks and spontaneous breech delivery of a premature stillborn female

infant followed. There was a post-partum haemorrhage of 40 oz. (1,140 ml.). Manual removal of the placenta was carried out. Post-mortem examination of the foetus showed multiple congenital abnormalities, including a type C congenital abnormality of the uterus.

Case 9.—Aged 41. Type B. Diagnosis: Two cervices were found at booking clinic. There was no vaginal septum. First pregnancy, 1954: This was in the right horn. The antenatal period was normal. Forceps delivery took place at 39 weeks' gestation. The indication was foetal distress. The infant weighed 6 lb. 1 oz. (2,750 g.). There was a post-partum haemorrhage of 21 oz. (600 ml.). The puerperium was not morbid.

Summary

Nine cases of pregnancy associated with congenital uterine abnormality are reported and a simple working classification is suggested.

The incidence of the condition is probably 1 in 1,000 pregnancies.

It is often associated with other congenital abnormalities, particularly of the urinary tract. For this reason a full urinary investigation should be done in all cases.

In types A, B, and D there is little extra risk to mother or foetus.

In types C, E, and F the maternal and foetal risks are maximal, owing to the increased incidence of abortion, malpresentation, and manipulations during labour.

The ante-partum complications and the complications occurring during labour are mentioned.

The place of elective caesarean section is discussed. It is suggested that its use be governed by the type of abnormality present, and only in types C, E, and F should elective caesarean section be considered.

I thank Mr. K. G. Seager, of Farnborough Hospital, and Miss I. R. Bishop, of St. James' Hospital, for permission to publish the cases, and Mr. G. M. Evans for help with the text.

REFERENCES

- Baker, W. S., Roy, R. L., Bancroft, C. E., McGaughey, H., Dickman, F. N., and Tucker, G. W. (1953). *Amer. J. Obstet. Gynec.*, **66**, 580.
 Berkeley, C., Bonney, V., and MacLeod, D. (1938). *The Abnormal in Obstetrics*, p. 200. London.
 Burton-Browne, J. R. C. (1948). *J. Obstet. Gynaec. Brit. Emp.*, **55**, 418.
 Corbet, R. M. (1946). *Irish J. med. Sci.*, p. 64.
 Dougal, D. (1921). *J. Obstet. Gynaec. Brit. Emp.*, **28**, 561.
 Fenton, A. N., and Singh, B. P. (1952). *Amer. J. Obstet. Gynec.*, **63**, 744.
 Hoffman, E. S. (1946). *Ibid.*, **51**, 692.
 Hunter, W. (1950). *J. Obstet. Gynaec. Brit. Emp.*, **57**, 721.
 Jarcho, J. (1946). *Amer. J. Surg.*, **71**, 106.
 Keevil, N. L. (1943). *J. Obstet. Gynaec. Brit. Emp.*, **50**, 146.
 Kerr, J. M., and Moir, C. (1949). *Operative Obstetrics*, p. 448. London.
 Kussmaul (1859). Quoted by W. Blair-Bell (1909). *Proc. roy. Soc. Med. (Obstet. and Gynaec. Sect.)*, **2**, 311.
 Mauriceau, F. (1675). *Traité des Maladies des Femmes*. Grones, Paris.
 Miller, N. F. (1922). *Amer. J. Obstet. Gynec.*, **4**, 398.
 Moore, Eliz. M. (1936). *J. Obstet. Gynaec. Brit. Emp.*, **43**, 936.
 Smith, F. R. (1931). *Amer. J. Obstet. Gynec.*, **22**, 714.
 Way, S. (1945). *J. Obstet. Gynaec. Brit. Emp.*, **52**, 325.
 Williams, B., and Cummings, G. (1953). *Ibid.*, **60**, 319.

A new single-story chest clinic at Chester City Hospital was formally opened in April by Mr. H. MORRISTON DAVIES, adviser in thoracic surgery to the Liverpool Regional Hospital Board. The clinic cost £12,800 to build and will serve the Chester and district hospitals area, which includes part of Flintshire. Mr. Morriston Davies pointed out that, before the Regional Board took over, the chest clinic was inadequately housed, with no beds of its own, no laboratory, and no operating theatre. The new clinic, though small—a big one was not needed—was magnificent, he said.