mild congestive phenomena. In such cases it appears justifiable to rely upon physiotherapeutic measures while in bed, and early ambulation, to prevent venous thromboembolism. In the case of older patients with severe congestive failure, however, oral anticoagulant drug therapy can be expected to prevent venous thromboembolism after the second week, and such prophylaxis is clearly indicated if its possible early thrombogenic tendency can be overcome. Concomitant high-dose intravenous heparin therapy during the first week or 10 days may surmount this difficulty, but, with the methods at present in use, would seriously interfere with the prothrombin estimations necessary for accurate adjustment of the dose of oral anticoagulant. Rapid attainment of therapeutically desirable prothrombin levels by means of heavier initial loading doses of a quicker acting oral anticoagulant such as ethyl biscoumarin-ace- late (Weiner et al., 1955), perhaps with heparin for the first day or two, may offer a preferable alternative.

Summary

The incidence of pulmonary embolism in 76 patients with congestive cardiac failure given prophylactic (oral) anticoagulant therapy was 8%. The incidence in a similar control series of 80 such patients not given anticoagulants was 10%.

Anticoagulant prophylaxis with phenindione, instituted at the time of admission, appears to afford protection against venous thromboembolic complications after the first 10 days, but this advantage is counterbalanced by an apparently increased tendency to such episodes during the first week or 10 days of administration of the drug.

The risk of thromboembolism in patients with clinically mild degrees of failure is too small to warrant routine anticoagulant drug prophylaxis.

We gratefully acknowledge the keen interest taken by the late Dr. O. Brenner in the establishment of this trial and his kind permission to study patients under his care. Our thanks are due also to Dr. W. C. Smallwood for his helpful criticism and advice, and to Mrs. P. Moffat for her patient secretarial assistance.

References


Pygopagus Conjoined Twins

J. M. GUPTA,* M.B., M.R.C.P.ED., D.C.H.

The occurrence of conjoined twins is rare. The incidence, according to Potter (1961) and Puegh (Guinard, 1893), is one in 50,000 births. Robertson (1953), in an analysis of 117 cases collected from the literature by Taruffi (1891-4), determined the sites of union. In 73% of the cases the junction was in the thoracic region (thoracopagus), in 19% it involved the sacral region (pygopagus), in 6% the pelvic outlet (ischio-pagus), and in 2% the head (craniopagus). Though there are numerous detailed reports of the separation of craniopagus and thoracopagus conjoined twins, no account of the separation of pygopagus and ischiopagus conjoined twins is to be found in the medical literature.

Case Report

These female conjoined twins (Joan and Jennifer) were self-delivered at home on 24 December 1961 to a 25-year-old Nigerian woman who had five other children. The mother stated that the first twin was born by the head, followed by the four legs and then the head of the second baby. After delivery the mother's general condition was good and her perineum was intact. There was a history of twinning in the father's family. The combined weight of the twins at birth was 3,640 g. During their stay in Nigeria, Jennifer (the left-sided twin) had a severe chest infection, and frequent watery stools were passed from the common anus. The twins were vaccinated separately on 21 March 1962, but as the vaccination did not take in Jennifer she was revaccinated on 26 March, when it took satisfactorily. They were transferred to Hammersmith Hospital on 6 April under the care of Professor Ian Aird.

The twins were joined at the buttocks, there being complete fusion of the left buttock of one with the right buttock of the other and incomplete fusion of the other buttocks, so that they were not completely back to back but could turn their heads to look at each other. Joan was on Jennifer's right-hand side (Figs. 1 and 2). The anal opening was common to the twins, but it was situated more to Joan’s side. The external genitalia were separate.

On admission to Hammersmith Hospital Joan was observed to be more lively and active than her sister. She had a grade 3/6 pansystolic murmur over the third and fourth left intercostal spaces paraesternally. Lungs and abdomen showed no abnormality. Cervical, thoracic, and upper lumbar spine were normal except for

*Late Neonatal Registrar, Hammersmith Hospital, London. Address from 1 August 1966: Department of Paediatrics, University of Khurum, Khurum, Sudan.
l lordosis. The lower lumbar spine and sacrum were fused to those of the other twin. She was alert, followed objects with her eyes and also by turning her head. She had a social smile and was able to hold her head up without support. Her hearing appeared to be normal. Deep reflexes and motor power were normal. On testing sensation to a prick in areas innervated by all the sacral segments of the spinal cord it was found that if one twin was pricked with a pin both would cry. The crying was not in sympathy, as this did not happen if they were pricked over other parts of their bodies, when only the infant pricked would cry. This observation suggested that the sacral segments of the twins conducted sensation to a common spinal cord or to spinal cords of both twins.

Jennifer's behaviour was independent of her sister's—for instance, when crying she was not soothed if her sister was quietened. Her sleep rhythm, movements of head, neck, arms, and legs, breathing, micturition, and social response were also independent of those of her sister. She was cyanosed and the peripheral pulses could not be felt. Her apex beat was not palpable; the heart sounds were best heard on the right side of the chest. There was a short soft (grade 2/6) mid-systolic murmur over the second and third left and right intercostal spaces parasternally. The lungs and abdomen were normal. The perineum showed a dimple with a sinus in the anal region which admitted a soft rubber catheter up to an inch (2.5 cm.).

Investigations

Blood groups of both twins were B Rh positive. X-ray examination of the lumbar spine (Fig. 3) showed that the twins were joined at the lower end of the sacrum. The sacra were defective in both twins and were joined end to end. There were five lumbar vertebrae in both spines, and one complete sacral segment, which appeared to be separate. The pelvic bones were separate and complete. Myelography (Fig. 4) demonstrated a common spinal theca with a small protrusion at the point of fusion. A spinal cord could be seen in each side, becoming narrow at the point of fusion. Barium enema (Fig. 5) demonstrated a common anal canal via which it was possible to fill the rectums and sigmoid colons of both infants. The rectums appeared to lie in a normal position relative to the anus. The sinus on the opposite side was filled with opaque material and was found to extend laterally and to end blindly. Joan's chest x-ray film showed a normal heart with clear lung fields. Because of the severe lordosis it was difficult to obtain a good x-ray picture of Jennifer's chest. In the course of repeated examinations the right hemithorax was found to be opaque, though the electrocardiogram did not suggest dextrocardia. Joan's electrocardiogram showed biventricular hypertrophy. The electroencephalograms were normal for their age.

![Fig. 2.— Twins before separation.](image)

![Fig. 3.— Radiograph of lumbar spine and sacrum, showing union at the sacrum.](image)

![Fig. 4.— Myelogram showing a common spinal theca with a small protrusion at the point of fusion.](image)

![Fig. 5.— Barium enema demonstrating the common anal canal.](image)
Progress

On admission both had diarrhoea. Culture grew Salmonella pamlouth and the infection was successfully treated with polymyxin and neomycin by mouth. On 15 May transverse colostomies were made as a preliminary in order to sterilize the guts distally. Next day Jennifer developed fever, signs of chest infection, and abdominal distension. She was treated with polymyxin, methicillin, and chloramphenicol. On 17 May she stopped breathing for 45 minutes, but there was no detectable change in her colour. She was quiet and lethargic and did not respond to painful stimuli; however, her eyes were open, and she appeared to be aware of her attendants, since she was looking around and following them with her eyes. During this time Joan started to breathe rapidly (rate 104/min.) though otherwise she was undisturbed. On pricking over the sacral segment of both twins with a pin Joan cried but Jennifer showed no response. Joan was given oxygen by face-mask, following which Jennifer started to breathe again. Jennifer continued to have short periods of apnoea over the next 24 hours, after which her condition improved; however, on 21 May her abdominal distension increased and the colostomy stopped functioning. This was followed by generalized abdominal tenderness and vomiting. Her condition continued to deteriorate during the next 12 hours. In the meantime Joan's respiratory and pulse rates had increased. On 22 May an emergency operation was performed by Professor Ian Aird and Mr. Geoffrey Knight, during the course of which Jennifer died.

Before the operation the twins were started on hydrocortisone (75 mg./day). They were anaesthetized separately. At the operation it was found that the two rectums were joined at an angle approximately 2 in. (5 cm.) from the anus. There was fusion of the spines below the fifth lumbar vertebrae. The lower spinal cord, the meninges in this region, and the cauda equina were common to the twins. During the operation it was obvious that Jennifer would not survive; hence the operation was so performed that the common anus, the common rectum, and the common spinal cord were left to Joan.

After the operation Joan continued to receive hydrocortisone, which was replaced by prednisolone after three weeks, the latter being discontinued on 23 June. She had a continuous leak of cerebrospinal fluid through a sinus. On 2 June the sinus was explored and an opening in the dura was repaired, following which the leak ceased. A plastic operation was undertaken on 2 August to refashion the right buttock where there was excessive tissue (Figs. 6 and 7). The colostomy was closed on 2 October and normal bowel action was established by 14 October.

On discharge from Hammersmith Hospital on 8 December 1962, she appeared a little backward for her age but there was no gross mental deficiency. She took a lively social interest in her surroundings, played with her toys, and had a vocabulary of half a dozen words. Though she could sit unsupported she was unable to sit herself up. The muscle tone, motor power, reflexes, and movements were normal in all the limbs. There were present a slight protuberant mass on the right buttock, lumbar lordosis, and lumbar scoliosis with the convexity to the right. The pelvis was tilted with apparent shortening of the right leg. The right hip clicked on abduction and x-ray examination showed subluxation with a shallow acetabulum and a small femoral epiphysis. The anus lay more to the right and the vulva lay obliquely across the median plane, running from left to right from above downwards. The pansystolic murmur was unchanged. She had a persistent anaemia, and investigations suggested that she had Hb/S trait. Professor R. G. Hendrie reported that at the age of 3 years she appeared to be completely normal mentally and there were no neurological abnormalities. The scoliosis, though still present, is slight and she walks with a limp.

Necropsy Findings in Jennifer (Abridged)

The body measured 48 cm. from crown to heel and weighed 3,500 g. The heart weighed 85 g. complete with the great vessels. The left ventricle measured 2.5 cm. in thickness. The right ventricle was hypertrophied and its thickness was 5.5 mm. The mitral orifice was 14 mm. in diameter, the tricuspid 15 mm., the pulmonary 9 mm., and the aortic 10 mm. All the heart valves appeared normal. There was no fibroelastosis and no obstruction of the right ventricular outflow tract. The coronary arteries were normal in position and size. The right atrium was a little distended. The aorta and its branches were normal. The pulmonary-artery trunk showed a little dilatation, measuring 10 mm. diameter, and the right pulmonary artery branched early. The ductus arteriosus was closed. There was only one pulmonary vein on each side. The systemic venous return was by way of a normal inferior vena cava and two superior vena cavae. The right superior vena cava entered the right atrium normally, but the left entered the coronary sinus. The foramen ovale was open but appeared valvular and measured 0.6 cm. in diameter. There was no abnormality in the heart to account for the right ventricular hypertrophy.

Each lung weighed 27 g. They were hyperplastic, sank in water, and appeared unerupted. The bronchi were normal.

In the alimentary system all organs appeared normal, except the rectum, which was balloononed through a deficiency in the posterior...
of O'Connell (1953), that to administer predicted and operation.

The cause of conjoined twins is not clear. Aird (1959) suggested that, as both twins are of the same sex and the union is at similar parts of their external surfaces, the majority are monozygotic. So far all conjoined twins have been found to have the same blood-group. Hamilton (1954) has suggested that the initial fault may be in the division of the blastocyst, which thereafter remains incomplete. It is possible that separate twins may become joined from fusion in the early stages of their development, but this would require the reappearance of the intervening amnion. Aird (1959) states that conjoined twins differ from each other in size and appearance and are never so identical as are separate monozygotic twins. Moreover, though the union is at similar parts of the external surfaces it is seldom symmetrical.

The history of separation of conjoined twins has been reviewed (Aird, 1954, 1959, B.M.J., 1964). There are records of attempts at operation on conjoined twins as far back as the sixteenth century (Münster, 1550). The first successful operation for thoracopagus was carried out in 1912 (Aird, 1954), for craniopagus in 1953 (Grossman et al., 1953), and for pygopagus in 1955 (Wilson, 1962). The success of the operation would of course depend on the extent of the union of the vital internal organs and on the interdependence of the twins. The problems of separation in conjoined twins are numerous, and in the case where it is obvious that one of the twins will definitely die during the process of separation ethical and legal considerations have to be taken into account. Some of these problems have been discussed by Robertson (1953), O'Connell (1964), and Franklin (1964). It is important that an accurate diagnosis of the extent of union should be made pre-operatively. In our case it was possible to predict the union of the spinal cords from the clinical examination, and this was confirmed by myelography and at operation.

There have been reports in the literature of sudden onset of shock in one of the twins after separation of conjoined twins (Aird, 1954). It is suggested that in such a case one of the twins is responsible for the greater part of the production of cortisone for the pair and that cortisone be administered to withstand the shock of the operation. It was for this reason that hydrocortisone was given, prophylactically before operation, to the twins and continued after operation in the surviving twin.

It is important to note in this case that, though Joan had been vaccinated successfully, vaccination took satisfactorily later in Jennifer (after the initial failure). This means that the successful vaccination in Joan had conferred no immunity to Jennifer and suggests that the humoral mechanisms in the twins were separate.

Jennifer's heart was grossly overweight. This was mainly due to the enlarged and hypertrophied right ventricle. No anatomical lesion was found in the heart to account for these changes. As the lungs were hypoplastic, the hypertrophy of the right ventricle may have been secondary to the lung lesion. The lung findings suggested that the infant could have led an independent existence and was probably being kept alive by her sister while they were joined. Further evidence for this view was available clinically when she stopped breathing for various periods without immediate ill effects.

Though the weight of the brain was small in relation to the age of the twins that died, it is not abnormal if one compares it with the brains of infants of the same weight. The main pathological findings were those of cerebral anoxia of recent origin with some evidence also of chronic anoxia. These changes may have been the result of the apneic periods that the infant had during the week before death.

**Summary**

A case of pygopagus conjoined twins is described. It was possible accurately to determine the extent of union of the spinal cord by a clinical examination. The aetiology of conjoined twinning is discussed, and the history of attempts at separation of conjoined twins is briefly reviewed.

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**References**


