A CASE OF CONJOINED TWINS

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Among 56,249 maternities in the Collaborative Perinatal Project, 615 were twin maternities. One of these resulted in stillborn thoracoomphalopagus MZ female twins with multiple cardiovascular, alimentary and other malformations. The case is of further interst in that the mother had a surgically removed brain tumor in childhood and exhibited neurological symptoms and bizarre behavior before and during pregnancy. Drugs and treatments which she received during pregnancy are not known to be teratogenic.

The case for this report has been drawn from the United States Collaborative Perinatal Project *, a prospective study of factors which affect the parents before and during pregnancy and their relation to the outcome of pregnancy. Among 56, 249 maternities in the Collaborative Perinatal Project over a period of six years, 615 were twin maternities. One of these resulted in stillborn thoracoomphalopagus MZ female twins with multiple cardiovascular, alimentary and other malformations. The frequency of conjoined twins appears to range between 1 in 50,000 to 1 in 100,000 births (Potter 1961, Bulmer 1970). Our data seem to agree with these estimates though it is unwise to draw conclusions from one case. The twins were born to a Negro mother at the Medical College of Virginia, and if we were observing only Negro births we would conclude that the frequency of conjoined twins is 1 in 26,000 births; while if we confined our observation to births which occurred at the Medical College of Virginia, one of the collaborating Institutions, we would conclude that the frequency of conjoined twins is 1 in about 5,000 births!

Be that as it may, the case is interesting because of the medical history of the mother before and during pregnancy, which may or may not be significant in the occurrence of this malformation.

The mother is a 19-year old Negro woman of low socioeconomic background. At age 10 she was hospitalized with complaints of severe occipital headaches, staggering gait, frequent falls, stiffness of the neck and double vision. The diagnosis of cerebellar tumor was made and at operation a cerebellar astrocytoma was removed. She had a rather stormy postoperative course with temperature elevation, meningismus and opisthotonos and had several lumbar punctures to relieve sterile meningitis. During the course of radiation therapy she received a total of 2,250 roentgen units.

She remained asymptomatic through adolesence though during this period she was generally shuggish, her thoughts and behavior were disorganized, she had visual hallucinations and episodes of dizziness and blackouts. She managed, however, to find employement, working in tobacco fields and as kitchen aid and waitress. She married, and at age 18 gave birth to a normal child who died at one month of age of pneumonia.

About two months before the last menstrual period of the study pregnancy she developed severe back pains,

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occipital headaches and diplopia which persisted when closing the one eye. Examination revealed intact cranial nerves, no pathological reflexes, normal cerebellar function tests, normal gait and Romberg, and full function of extraocular movements. The symptoms persisted throughout her pregnancy and she received Darvon, 65 mg q.i.d. for relief of pain.

One month before her last menstrual period her bizzarre behavior became more pronunced, she complained of a knocking feeling in her head, ringing in the ears and vertigo. After consumption of alcohol she went into a psychotic episode for which she was hospitalized for three days. On psychiatric evaluation the diagnosis of acute anxiety and postoperative chronic brain syndrome was made. She was treated for brief periods of time with thorazine, reserpine, Na luminal, nembutal, phenobarbital and dilantin. During pregnancy she was also treated with achromycin, streptomycin and furacin for chronic cervicitis and received the usual supplements of iron and vitamins.

Shortly before delivery she was admitted to the hospital with mild preeclampsia, which is not uncommon with twin pregnancies. After several days in the hospital without specific response to therapy for preeclampsia she went into labor and at 30 weeks gestation she delivered spontaneously and without difficulty by breech presentation a female set of twins, joined by skin and bone from the area of the umbilicus to approximately the area of the second rib (Fig. 1).

The babies were grossly well formed though there was purplish discoloration, maceration and edema of body tissues. Together they weighed 1900 g and each measured 34.5 cm in leght. There was a common umbilical cord (Fig. 2) measuring 53 cm in length and containing six vessels. The cord divided at the placenta and the two branches inserted approximately 4 cm apart. The single placenta was large and macerated but grossly normal. One of the twins had right athelia.

At autopsy the twins were found to have fused hearts and fused livers. The heart essentially consisted of three chambers (Fig. 3) one of which contained elements of four rudimentary, separated atria. The tricuspid valve was absent. The pulmonary veins on twin A's side were absent and drainage went via a common vein to the portal region. On twin B's side there was a single draining pulmonary vein.

The right ventricle of twin B was a large, blind sack with pulmonary atresia and absent ductus arteriosus, fused to the left ventricle of twin A. The effective circulation was from the common auricle to a large common ventricle giving rise to a separate aorta and pulmonary artery for twin A and an aorta for twin B. A right subclavian artery arising from the left ringed the trachea of twin A.

The livers and the umbilical cords were fused.

Other gross and microscopic findings were compatible with the gestational age of the fetuses and the cardiovascular malformations; otherwise they were unremarkable. The umbilical cord had two bundles of three vessels each and the placenta was monoamniotic monochorionic.

The embryology of conjoined twins is not certainly known. Some believe that two centers of axial growth, instead of one, develop on the embryonic disc and if these are not sufficiently separated, they result in the sharing of the intermediate area by the two embryos. Others hold the view that the original embryonic disc undergoes incomplete fission and that the degree and origin of the splitting determine the variety of type of conjoined twins.

The cause of conjoined twinning is unknown. Witschi (1934) thought that delayed ovulation leading to ageing of the ovum could result in gradual decline and inability of the ovum to differentiate normally and that two centers of organization, neither able to suppress the other, result. It is difficult to assess factors predisposing to ageing of the ovum in this case. Contraception has been suggested as one such factor but the mother stated that since the death of her first child she had heen trying to have another child and that neither she nor her husband used contraceptive devices. She did, however, have chronic cervicitis during pregnancy, which conceivably might have had adverse effects on the ovum but it is not known how early in pregnancy the infection began.

The condition is supposed to be determined by the end of the second week of gestation which makes the influence of teratogenic agents extremely difficult to assess. Though the mother was treated with several drugs, most of them were given only once and we could not trace a responsible teratogenic agent which was given during the critical period. The mother started taking Darvon (dextropropoxyphene HCl) for pain about two months before conception and, presumably, during the critical early gestational period but this drug has not been previously implicated in teratogenesis.

The mother had two pregnancies subsequent to that of the conjoined twins and, as far as we have been able to ascertain, these resulted in liveborn children without obvious malformations.

Fig. 1. Stillborn thoracoomphalopagus twins.

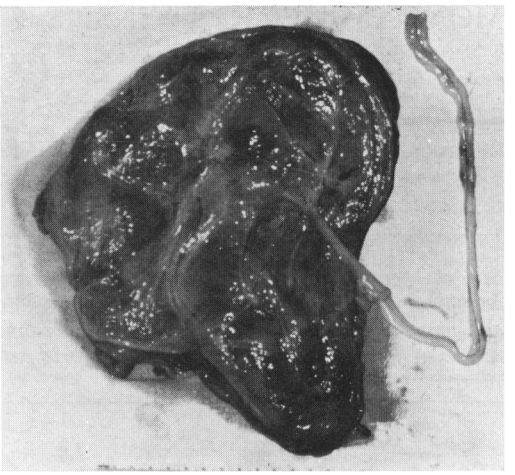

Fig. 2. The placenta with common (fused) umbilical cord.

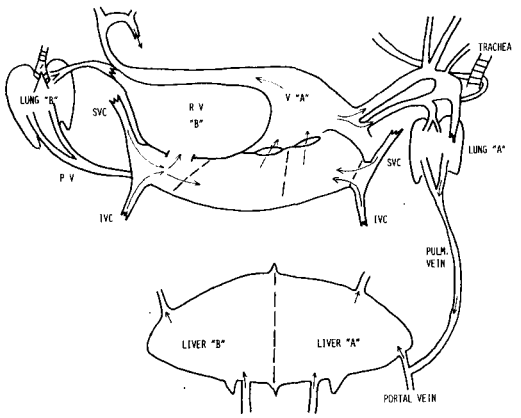

Fig. 3. Diagram of the cardiovascular malformations and effective circulation of the twins.

REFERENCES

Bulmer M.G. 1970. The Biology of Twinning in Man. Oxford: Clarendon Press.

Potter E.L. 1961. Pathology of the Fetus and Infant [2nd edition]. Chicago: Year Book Medical Publishers Inc.

Witschi E. 1934. Appearance of accessory "organizers" in overripe eggs of the frog. Proc. Soc. Exp. Biol. Med., 31: 419.

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