ARTERIOVENOUS ANEURYSM OF THE GREAT VEIN OF GALEN WITH HEART FAILURE IN THE NEONATAL PERIOD

BY

A. E. CLAIREAUX* and C. G. H. NEWMAN

*From the Institute of Obstetrics and Gynaecology, and the Institute of Child Health,
University of London*

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The great cerebral vein of Galen extends from the junction of the two internal cerebral veins under the splenium of the corpus callosum to the commencement of the straight sinus. Arteriovenous aneurysms of this vein are rare congenital anomalies and only 22 cases have been found in a review of the literature.

Other types of cerebral arteriovenous aneurysms are not so unusual. Their true incidence has become more apparent following the widespread use of angiography. Mackenzie (1953) estimated their occurrence as 1% of all neurological admissions. By way of contrast Paterson and McKissock (1956) found no example of great cerebral vein aneurysm in 110 cases of cerebral arteriovenous aneurysm, and the same is true of the series reported by Olivecrona and Riives (1948), Mackenzie (1953) and Potter (1955). These vascular malformations are predominantly of two main types (Hamby, 1952). In the first type, there is a direct end-to-end connexion between an artery and vein, and in the second a network of poorly differentiated, non-capillary vessels intervenes between an artery and several veins, forming a mass of abnormal, dilated and tortuous vessels which sometimes goes by the name of 'angioma' but is not generally considered to be neoplastic. In both types veins not directly involved in the malformation may become enlarged and tortuous in response to the raised venous pressure. The great cerebral vein, however, only shows aneurysmal dilatation when there are one or more abnormal arterial vessels opening into it, its immediate tributaries or continuation. Other vascular malformations of the brain may coexist. These abnormal arterial vessels have arisen, in the described cases, from the posterior cerebral, posterior communicating, callosal and superior cere-

bellar arteries and from the choroid plexus of the third ventricle.

Local and general effects may be exerted on the cardiovascular system as in other varieties of arteriovenous aneurysm. The general effects may be difficult to detect. They include a variable increase in pulse pressure, tachycardia, apical systolic murmur and evidence of some enlargement of the heart. The literature contains reports of two babies who died of heart failure associated with a cerebral arteriovenous aneurysm not involving the great cerebral vein (Silverman, Breckx, Craig and Nadas, 1955) (Table 2) and three cases associated with an arteriovenous aneurysm of that vein (Table 1, Cases 11, 13 and 14). One of these infants had coarctation of the aorta as well.

The two cases reported here also died of heart failure associated with arteriovenous aneurysm of the great vein of Galen.

Case Reports

Case 1. S.F. was born September 15, 1959, by normal delivery at about 38 weeks' gestation. The birth weight was 2,240 g. and the head circumference was 33 cm. Respiration was established immediately.

Lower rib recession was noted 16 hours after birth and became more severe. The baby's extremities became cyanosed, peripheral pulsations were poor, there was a soft systolic murmur and the liver became enlarged. The respiratory rate rose to 70 per minute. This picture of respiratory distress and early heart failure was complicated by the occurrence of head retraction and opisthotonus 30 hours after birth followed by twitching and frank fits. Fontanelle tension was normal. Blood calcium was normal. Blood glucose levels were persistently low, 23 and 15 mg. % in the first 24 hours and never above 15 mg. % subsequently in spite of hourly glucose feeds. Lumbar puncture showed xanthochromic fluid under moderately increased pressure with 14,700 red and 75 white blood cells per cu. mm.

* Present address: Queen Charlotte's Hospital, London W.6.

Author	Case No.	Lesion and Arterial Supply	Presenting Symptoms or Signs
Steinheil (1895)	1	Left callosal artery; great and one small cerebral vein dilated; sacculated aneurysm on pedicle on great cerebral vein. Associated right frontal vascular malformation	Convulsions; left hemianopia
Jaeger, Forbes and Dandy (1937) Jaeger and Forbes (1946)	2	Huge dilated great cerebral vein pressing on aqueduct; large branches off both post. cerebral arteries into small cerebral veins; further vascular malformations	Internal hydroceph.; epistaxis, anaemia
Russell and Nevin (1940)	3	Atresia of int. jugular veins. Left post. cerebral artery branch to great cerebral vein; serpentine mass of vessels	Prominent face and neck veins; arrested motor development; exophthalmos; cranial bruit
Russell and Nevin (1940)	4	Sup. callosal art. and post. cerebral arteries, branches to great cerebral vein; aneurysm compressed aqueduct	Hydrocephalus; distended scalp veins
Alpers and Forster (1945)	5	Choroidal artery and post. cerebral arteries, branches to aneurysm of great cerebral vein	Mental retardation CNS signs; internal hydrocephalus
Oscherwitz and Davidoff (1947)	6	Tumour probably represented aneurysm of great vein of Galen and branches of circle of Willis	Headaches
Lumsden (1947)	7	Aneurysm on upper surface of tentorium attached to near great vein of Galen at junction of inf. sagitt. sinus and straight sinus; ? small artery from choroid plexus of 3rd ventricle	Fits; intraventric. and subarach. haemorrhage
Boldrey and Miller (1949)	8	Branches of post. cerebral artery to aneurysm of great cerebral vein; pulsatile mass of vessels behind right mastoid	Hydrocephalus; dilated face, scalp veins; pulsatile mass behind mastoid; repeated subarach. haemorrhage
Boldrey and Miller (1949)	9	Post. communic. and post. cerebral arteries connected to great cerebral vein (angiogram)	Swollen face, eye veins; fatigability
Wolfe and France (1949)	10	'At least one artery entered aneurysm of great vein of Galen'	Fits; CNS signs; int. hydrocephalus
Pollock and Laslett (1953)	11	Post. cerebral and sup. cerebral arteries communic. with aneurysm of great vein of Galen and sag. sinus; also vessels from int. cerebral veins, most of supply vessels appeared to be thrombosed	Heart failure; large head
Petit-Dutaillis, Guiot and Berdet (1953)	12	Branches of post. cerebral arteries to aneurysm of great vein of Galen	Hydrocephalus
Clément, Gerbeaux, Combes-Hamelle, Pertuiset and Petranca (1954)	13	Feeding vessels to aneurysm of great cerebral vein not identified; much thrombus	Dyspnoea; increased muscular tone; dysphagia; cyanotic attacks
Clément <i>et al.</i> (1954)	14	Post. cerebral arteries communicate with aneurysm of great cerebral vein; also 'very abnormal arterial distribution'	Failure to thrive; wasting; hydroceph.; dilated veins; cranial bruit; dyspnoea
Clément <i>et al.</i> (1954)	15	Personal communication on similar case	No details
French and Peyton (1954)	16	Anterior cerebral artery group communic. with great cerebral vein	Large head; failure to thrive; mental retardation
French and Peyton (1954)	17	Abnormal mass of vessels on medial side. Right cerebral hemisphere, communic. with great cerebral vein, ant. callosal artery enlarged	Sub-arach. haemorrhages
French and Peyton (1954)	18	Arteriovenous malformation near pineal with rapid filling of great vein of Galen; abnormally large ? left ant. cerebral artery	Headache; slurred speech
French and Peyton (1954)	19	Branches of post. cerebral arteries into much dilated great vein of Galen	Headaches; subarach. haemorrhages
French and Peyton (1954)	20	Enlarged sup. great cerebral artery into large aneurysmal sac near pineal, draining into dilated great vein of Galen	Headaches; photophobia; sweating, lethargy; subarach. haemorrhage; mental illness
Hirano and Terry (1958)	21	Branches of post. cerebral arteries seemed to end blindly in wall of aneurysm of great cerebral vein	Hydrocephalus, epistaxis, proptosis, pyrexia, ? cause
Gibson, <i>et al.</i> (1959)	22	Branches of post. cerebral artery to aneurysm of great cerebral vein, compressing aqueduct	Hydrocephalus
Claireaux and Newman (Case 1)	23	Arteriovenous aneurysm of both post. cerebral arteries and great cerebral vein	Heart failure, fits, opisthotonos
Claireaux and Newman (Case 2)	24	Arteriovenous aneurysm of right post. cerebral artery and great cerebral vein	Neck retraction

OF THE GREAT CEREBRAL VEIN OF GALEN

Age at Onset	Age at Diagnosis	Associated Cardiovascular Signs	Surgery	Outcome
Child	49 yrs	—	—	Died
8 mths	4 yrs	Murmur and increased cardiac dullness after 'flu at 8 mths; bruit in neck at 27 mths, low blood pressure, anaemia; carotids dilated, also jugulars	Carotid ligation	Died
3 mths	17 mths	—	—	Died
2 mths	17 mths	None in life	Decompression	Died
6 yrs	18 yrs	—	Craniotomy	Died
26 yrs	27 yrs	Soft blowing systolic murmur	Craniotomy	Alive
4 mths	7 mths	—	—	Died
8 mths	16 mths	—	Carotid ligation; clipping of feeding arteries	Alive
Birth	15 yrs	—	Carotid ligation	Alive
3 yrs	21 yrs	—	Torkildsen's operation	Died
Birth	4 days	Heart failure	—	Died
4 mths	4 mths	—	—	Alive
? Birth 10 days	1 mth	Heart failure	—	Died
? 3 wks	7 mths	Heart enlarged; coarctation of aorta	—	Died
Infancy	—	—	—	Died
2 mths	21 mths	—	Craniotomies; clipping of feeding arteries	Alive
?	12 yrs	—	Clipping of feeding arteries	Alive
?	22 yrs	—	Clipping of feeding arteries	Alive
?	14 yrs	—	—	Alive
?	43 yrs	—	Craniotomy	Alive
5 mths	28 mths	—	—	Died
6 mths	4½ yrs	—	Anastomosis of spinal theca to peritoneum	Died
16 hrs	4 days	Cyanosis, systolic murmur, neck pulsations, heart failure	—	Died
24 hrs	16 days	Cyanosis, systolic murmur, neck pulsations	—	Died

Note:—Nil or not mentioned

TABLE 2
HEART FAILURE ASSOCIATED WITH CEREBRAL ARTERIOVENOUS ANEURYSM NOT INVOLVING GREAT CEREBRAL VEIN OF GALEN

Author	Case No.	Lesion and Arterial Supply	Presenting Symptoms or Signs	Age at Onset	Age at Diagnosis	Associated Cardiovascular Signs	Surgery	Outcome
Silverman <i>et al.</i> (1955)	1	Right cerebral hemisphere almost replaced by massive vascular network penetrating right frontal bone; right middle and post. cerebral arteries gave branches to communicate with dilated sup. sagitt. sinus	Dyspnoea; cyanosis	4 hrs	1 day	Systolic murmur; right carotid thrill; cardiac enlargement and failure	—	Died
Silverman <i>et al.</i> (1955)	2	Large cirroid aneurysm between middle cerebral artery and sigmoid sinus; large tortuous vascular channels	Respiratory distress; cyanosis; shrill cry	30 hrs	3 days	Systolic murmur; venous congestion; cardiac enlargement	—	Died

The cerebrospinal fluid glucose level was 30 mg. %. The opisthotonus improved markedly after the lumbar puncture but relapsed. The fits, some left sided, recurred. Neck pulsations became very vigorous. Venous congestion, further enlargement of the liver and peripheral oedema developed. Digitalization caused only a transient and partial reversal of these signs. An electrocardiogram showed digitalis effect and increased P waves and some evidence of left as well as of right ventricular enlargement. The baby died on the fourth day, in heart failure. Blood glucose levels had shown no sign of rising.

ELECTROENCEPHALOGRAM. 'The pattern of this record is that of a full term normal baby when awake and sleeping. There is perhaps a little more rhythmic activity at 15 c./sec. than usual but this may be accounted for by the prematurity. This activity is mostly seen when the baby is awake and is up to 25 μ V in amplitude. When the baby is asleep there are repeated single or multiphasic sharp waves up to 250 μ V in amplitude and up to duration 0.5 sec. These slow sharp waves are unilateral and occur simultaneously over all areas of the head and about equally on both sides' (Dr. Ruth Harris).

RADIOLOGY. Skull radiograph revealed no abnormality; chest radiograph showed only moderate cardiac enlargement.

NECROPSY. The body was that of a small male infant weighing 2,150 g. The dura mater and pia arachnoid were healthy. The brain (333 g.) was firm. The torcular Herophili was greatly enlarged and contained firm clot. Clot was also present in the superior and inferior longitudinal sinuses, the transverse sinuses and the jugular bulbs. The jugular veins appeared to be normal. At the base of the brain a large aneurysm was found. This was largely composed of a grossly distended great cerebral vein of Galen which communicated anteriorly with both posterior cerebral arteries and posteriorly with the distended torcular (Fig. 1). The aneurysmal sac measured 2.2×1.5 cm. The arterial communication was complex. The basilar

artery divided anteriorly into the right and left posterior cerebral arteries in the usual manner. The right posterior cerebral artery then divided immediately into four and these reunited after passing downwards and backwards for a distance of 3 cm. The thick vessel thus formed then subdivided again before uniting with the aneurysm (Fig. 2). The left posterior cerebral artery divided into three and the branches joined the anterior part of the aneurysm. The great cerebral vein after receiving the internal cerebral veins expanded into a thick-walled sac. This sac received both posterior cerebral arteries and these passed backwards to join the torcular Herophili. The aneurysm had a bulbous anterior portion, a narrow neck and a fusiform body. The cerebral veins were not distended. The lower portion of the

FIG. 1.—Case 1, brain; the inferior surface is exposed to show the aneurysm of the great cerebral vein and the abnormal communications with the posterior cerebral arteries.

aqueduct of Sylvius was of normal calibre and the fourth ventricle was not distended. The anterior surface of the cerebellum was hollowed in the region of the aneurysm. The brain was congested.

The pericardium was clear and glistening and there was a moderate quantity of clear, yellow fluid in the pericardial sac. The heart was enlarged as a result of dilatation of the right atrium and right ventricle. Both chambers contained a large amount of blood clot. The foramen ovale and ductus arteriosus were patent. The valves and great vessels had a normal appearance.

The larynx, trachea and bronchi were healthy. The lungs were congested and some subpleural haemorrhages were present. On slicing a few haemorrhagic areas were noted. The stomach and bowel were healthy. The liver (52 g.) and spleen (5 g.) were not enlarged. The genito-urinary tract was healthy and the endocrine glands showed no abnormality. Supernumerary digits were present on the lateral border of each hand. There was a complete extra finger on the left hand and a rudimentary one on the right. The fifth toe of the right foot was bifid.

HISTOLOGY.

Lungs. The pleura was healthy. The lungs were congested but quite well expanded. Some patchy haemorrhage was present. In some areas the alveolar ducts and alveoli were flooded with red cells. The bronchi were healthy. There was no pneumonia.

Aneurysm. A section was taken through the most dilated portion of the great cerebral vein. The lumen contained fibrin thrombus. The wall was quite thin and there was evidence of degeneration. No intima was recognized.

Left Posterior Cerebral Artery. The lumen was

FIG. 2.—Case 1, brain; outline drawing to show (a) aneurysm of great cerebral vein, (b) right posterior cerebral artery with abnormal branching, (c) left posterior cerebral artery, (d) basilar artery.

occluded by fresh thrombus which showed no evidence of organization. The vessel was distended as a result of the enclosed thrombus. The wall showed no degenerative change.

SUMMARY. Cerebral arteriovenous aneurysm; intrapulmonary haemorrhage; supernumerary digits on each hand and bifid right fifth toe.

Case 2. The mother, 23 years of age, para 0 had bilateral active tuberculosis and during pregnancy had to be treated for pyelonephritis resulting from infection with *Escherichia coli*. She went into labour at term and the infant was delivered spontaneously. The first stage of labour lasted 18 hours 30 minutes and the second stage 30 minutes. There was a vertex presentation.

The infant was a male and weighed 3,715 g. at birth. The head circumference was 32.5 cm. The infant's condition was good and he cried immediately. There was no peripheral cyanosis but cardiac pulsation was noted to be vigorous and there was strong arterial pulsation in the neck. A systolic murmur was heard over the apex. On radiological examination the heart was seen to be enlarged. One week after birth cyanosis was observed and there was slight pitting oedema of the feet and legs. The pulse rate was 160 per minute and respirations 40 per minute. A diagnosis of congenital malformation of the heart was made. Further radiological examination on the 11th day showed an increase in the size of the heart. Cyanosis became more marked and neck retraction was noted. The infant's condition steadily deteriorated and he died 16 days after delivery.

NECROPSY. The body was that of a well developed male infant and weighed 3,345 g. The sternum was prominent. The falx and tentorium were intact. The brain (400 g.) was very soft and markedly congested. The great cerebral vein was replaced by a large saccular aneurysm 4.5 cm. in length and 2 cm. in diameter. This aneurysm was attached to the junction of the falx with the tentorium. It communicated anteriorly with the right and left internal cerebral veins and with the right posterior cerebral artery. The cerebral veins were engorged but not thrombosed. There was slight dilatation of the lateral ventricles and of the third ventricle which contained clear cerebrospinal fluid. The aqueduct was patent and the fourth ventricle was of normal size. A small amount of subarachnoid haemorrhage was present over the cerebellum. On slicing the aneurysmal dilatation of the great cerebral vein was found to contain organized thrombus.

The pericardium was clear and glistening, there was a small quantity of clear, straw-coloured fluid in the pericardial sac. The heart was considerably enlarged, chiefly as a result of hypertrophy and dilatation of the right atrium and right ventricle. The interventricular septum was intact and very broad. Thick post-mortem clot was present in the right atrium. There was no structural abnormality of the heart valves or of the great vessels. The foramen ovale was closed. The ductus

FIG. 3.—Case 2, heart; right ventricle opened to show gross myocardial hypertrophy.

arteriosus was narrowly patent. The venous drainage was normal (Fig. 3).

The larynx, trachea and bronchi were healthy. The lungs were moderately well expanded and congested. Some patchy basal consolidation was found.

The stomach and bowel showed no pathological change. The liver (149 g.) was enlarged and congested. The spleen (15 g.) was large and very congested. The genito-urinary tract was healthy. The endocrine glands showed no abnormality.

HISTOLOGY.

Lungs. The pleura was healthy. The lungs were congested and some haemorrhage had occurred into the alveoli. In sections from the lower lobes there were widespread areas of consolidation. Many alveoli and alveolar ducts contained a dense cellular exudate composed of polymorphonuclear leucocytes and histiocytes. Some small bronchi contained a similar exudate. Immediately beyond the consolidated areas there was a zone of overdistension of alveoli and alveolar ducts. The appearance was that of a bronchopneumonia.

SUMMARY. Arteriovenous aneurysm of great cerebral vein; hypertrophy and dilatation of the heart, and terminal bronchopneumonia.

Discussion

Aneurysm of the great vein of Galen differs clinically from the more common types of cerebral

arteriovenous aneurysm in the early onset of symptoms and the apparently greater frequency of serious cardiovascular effect. Moreover, hydrocephalus is a commonly associated feature probably as a result of pressure by the aneurysm on the aqueduct of Sylvius. Hydrocephalus was not present in either of our patients. This is probably the result of the comparatively short duration of any pressure effects. In both the brains we examined, the aqueduct was still widely patent although slightly distorted. Similarly, we found no thickening of the wall of the aneurysm and no evidence of mucinous degeneration such as was reported by Russell and Nevin (1940). This lack of degenerative change appears to indicate that the dilatation of the vein is not secondary to a degeneration of the vessel wall as these workers suggested. In some instances (Russell and Nevin (1940), Case 1) a genuine arteriovenous aneurysm and a vascular hamartoma are present together. In our Case 1 there was some very peculiar branching of each posterior cerebral artery before union with the great cerebral vein took place, but in neither case was there any evidence of a network of fine vessels which might be designated a hamartoma.

In the 24 cases listed (Table 1) symptoms had commenced in infancy or childhood in two, at less than 6 months in 12, at 6 months to 1 year in three, at 1 to 10 years in two and in adult life in only one case. The date of onset was not stated in four cases who were diagnosed at 12, 14, 22 and 43 years respectively. In the more usual type of cerebral arteriovenous aneurysm or 'angioma' symptoms are more usually delayed until the second decade or later (Olivecrona and Riives, 1948; McKissock, 1950; Mackenzie, 1953). Cases presenting in infancy or childhood are proportionately rare.

In great cerebral vein aneurysms the symptomatology includes dilated face and scalp veins, proptosis, fits, headache, the effects of subarachnoid and intraventricular haemorrhage, mental retardation, focal signs in the central nervous system and failure to thrive. General circulatory effects were noted in six of the 24 cases and heart failure appeared to be the primary cause of death in five of these. One had coexisting coarctation of the aorta. Hydrocephalus developed in nine cases, two more having a 'large head'.

The course at first sight appears to be short. Death occurred under the age of 2 in nine, under the age of 5 in three and at 18, 21 and 49 years in a further three. Only one of seven cases reported before 1947 and before angiography was performed for diagnosis was alive at the time of reporting. This contrasts with eight of the remaining patients

and suggests that the prognosis may not really be so gloomy. Death was associated with internal hydrocephalus, fits, haemorrhage, craniotomy and heart failure.

Diagnosis was made by angiography, at necropsy or at operation. An intracranial bruit was not listened for in the two cases reported here and, when sought in other cases, has only sometimes been detected. The association of local or general cardiovascular abnormalities with a suspected intracranial lesion is characteristic of these malformations. The retinal veins appeared normal in the two cases described here.

Treatment is medical and surgical. The former has little to offer. It cannot influence the aneurysm nor the shunt, and the treatment of associated heart failure has been quite unsuccessful. Surgery has had some successes. It was carried out in 10 cases. Carotid ligation, attempts to relieve hydrocephalus and simple craniotomy had poor results. Surviving patients had undergone the following procedures: Craniotomy only, two; carotid ligation, one; carotid ligation with subsequent clipping of arteries feeding the aneurysm, one; clipping of feeding arteries, three. French and Peyton (1954) consider the condition operable if the arterial supply to the aneurysm can be demonstrated by angiography. Occlusion of the feeding arteries (which appears to have taken place spontaneously in Case 21) may not influence associated hydrocephalus, but will probably prevent pressure effects from further enlargement. It must also reduce the likelihood of haemorrhage and should correct the effects of the shunt on the cerebral circulation and on the heart itself. The youngest patients in whom feeding arteries were clipped were both under 2 years of age. Both survived but neither had been in heart failure. It is doubtful whether any of the infants who died of this complication could have been saved, but it is just possible that carotid occlusion might have improved their circulatory status temporarily.

The cardiovascular effects associated with arteriovenous aneurysms may be extensive. They have been reviewed by Lewis and Drury (1923), Holman (1923, 1924), Reid (1925a, b), McGuire (1935) and Potter (1955). Several authors had noted cardiac enlargement in cases of cerebral arteriovenous aneurysm, but Schmitt (1920) appears to be the first to have stressed this feature. He wrote: 'Hypertrophy of the left ventricle . . . is often found in the more advanced stages of cerebral arteriovenous aneurysm' and 'the combination of brain disorder with cardiovascular symptoms is very important in the diagnosis of cerebral arteriovenous

aneurysm'. He further mentions the dilatation of arteries to the head. In subsequent accounts the effects on the cardiovascular system are not always mentioned. Their incidence is therefore difficult to assess. Cushing and Bailey (1928) and Dandy (1928) found definite cardiac enlargement lacking in their cases even though it might have been expected in some on the grounds of size of aneurysm, location and duration of symptoms. Brock and Dyke (1932) found cardiac enlargement in four of five cases of cerebral arteriovenous 'angioma'. Ray (1941) noted partly reversible cardiac enlargement in one of 16 cases. Shenkin, Spitz, Grant and Kety (1948) found it in two cases investigated to determine the cerebral blood flow, which was much increased. Silverman *et al.* (1955) record two infants who died in heart failure from this cause in the newborn period and Paterson and McKissock (1956) noted cardiac enlargement in nine of their series of 110 cases. Aneurysm of the great vein of Galen does not feature in these reports but enlargement has occurred in four reported cases, three of whom died in heart failure, as did the two additional babies reported here. The very marked effect on the cardiovascular system exerted by the arteriovenous shunt is indicated in Case 2 where there is enormous hypertrophy of the wall of the right ventricle which was three times the normal thickness for this stage of development.

Hypoglycaemia has not been previously described in this context. It may have been coincidental, but the proximity of the aneurysm to the floor of the fourth ventricle is suggestive.

The cases presented here and the review of the literature make it apparent that cerebral arteriovenous aneurysms may present with symptoms referable to the cardiovascular system. The possibility of such lesions should be considered when a vigorous cardiac impulse and increased neck pulsations are associated with signs of meningeal irritation or convulsions. Respiratory distress may mask these features in the newborn period. If it is possible to confirm the diagnosis by angiography, surgery may offer a chance of successful treatment. Untreated, the patient may die in the newborn period from cardiac failure, or subsequently from the effects of hydrocephalus and haemorrhage.

Summary

Two cases of arteriovenous aneurysm of the great vein of Galen are reported. Both were infants who died in the newborn period of heart failure. Both exhibited neck rigidity in addition to cardiovascular signs. One infant had persistent hypo-

glycaemia. Clinical features of this type of arteriovenous aneurysm are reviewed and it appears that hydrocephalus and severe cardiovascular effects are commoner than in the 'angioma' type of cerebral arteriovenous aneurysm. Surgery appears to hold out the possibility of cure in some cases, provided that diagnosis and angiography anticipate the potentially rapid decline of such patients.

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REFERENCES

- Alpers, B. J. and Forster, F. M. (1945). Arteriovenous aneurysm of great cerebral vein and arteries of circle of Willis. *Arch. Neurol. Psychiat. (Chicago)*, 54, 181.
- Boldrey, E. and Miller, E. R. (1949). Arteriovenous fistula (aneurysm) of the great cerebral vein (of Galen) and the circle of Willis. *Ibid.*, 62, 778.
- Brock, S. and Dyke, C. G. (1932). Venous and arterio-venous angiomas of the brain. A clinical and roentgenographic study of 8 cases. *Bull. neurol. Inst. N.Y.*, 2, 247.
- Clément, R., Gerbeaux, J., Combes-Hamelle, A., Pertuiset, B. and Petranca, C. (1954). Anévrysmes artério-veineux de l'ampoule de Galien chez le nourrisson. *Presse méd.*, 62, 658.
- Cushing, H. and Bailey, P. (1928). *Tumours Arising from the Blood-vessels of the Brain*. Thomas, Springfield, Illinois.
- Dandy, W. E. (1928). Arteriovenous aneurysm of the brain. *Arch. Surg. (Chicago)*, 17, 190.
- French, L. A. and Peyton, W. T. (1954). Vascular malformations in the region of the great vein of Galen. *J. Neurosurg.*, 11, 488.
- Gibson, J. B., Taylor, A. R. and Richardson, A. E. (1959). Congenital arteriovenous fistula with an aneurysm of the great cerebral vein and hydrocephalus treated surgically. *J. Neurol. Neurosurg. Psychiat.*, 22, 224.
- Glatt, B. S. and Rowe, R. D. (1960). Cerebral arteriovenous fistula associated with congestive heart failure in the newborn: Report of two cases. *Pediatrics*, 26, 596.
- Hamby, W. B. (1952). *Intracranial Aneurysms*. Thomas, Springfield, Illinois.
- Hirano, A. and Terry, R. D. (1958). Aneurysm of the vein of Galen. *J. Neuropath. exp. Neurol.*, 17, 424.
- Holman, E. (1923). The physiology of an arteriovenous fistula. *Arch. Surg. (Chicago)*, 7, 64.
- (1924). Experimental studies in arteriovenous fistulas. *Ibid.*, 9, 856.
- Jaeger, J. R. and Forbes, R. P. (1946). Bilateral congenital arteriovenous communications (aneurysm) of the cerebral vessels. *Arch. Neurol. Psychiat. (Chicago)*, 55, 591.
- , — and Dandy, W. E. (1937). Bilateral congenital cerebral arteriovenous communication aneurysm. *Trans. Amer. neurol. Ass.*, 63, 173.
- Lewis, T. and Drury, A. N. (1923). Observations relating to arteriovenous aneurysm. *Heart*, 10, 301.
- Lumsden, C. E. (1947). A case of aneurysm of the vein of Galen. *J. Path. Bact.*, 59, 328.
- Mackenzie, I. (1953). The clinical presentation of the cerebral angioma. *Brain*, 76, 184.
- McGuire, J. (1935). Circulatory studies on a case of arteriovenous aneurysm. *Amer. Heart J.*, 10, 360.
- McKissock, W. (1950). Intracranial angiomas. *Ann. roy. Coll. Surg. Engl.*, 1, 472.
- Olivecrona, H. and Riives, J. (1948). Arteriovenous aneurysms of the brain. *Arch. Neurol. Psychiat. (Chicago)*, 59, 567.
- Oscherwitz, D. and Davidoff, L. M. (1947). Midline calcified intracranial aneurysm between occipital lobes. *J. Neurosurg.*, 4, 539.
- Paterson, J. H. and McKissock, W. (1956). A clinical survey of intracranial angiomas with special reference to their mode of progression and surgical treatment. A report of 110 cases. *Brain*, 79, 233.
- Petit-Dutaillis, D., Guiot, G. and Berdet, H. (1953). Anévrysmes cirsoïdes et anévrysmes artério-veineux de l'encéphale d'après dix-neuf cas considérations anatomiques, cliniques et radiologiques. *Presse méd.*, 61, 901.
- Pollock, A. Q. and Laslett, P. A. (1953). Cerebral arteriovenous fistula producing cardiac failure in the newborn infant. *J. Pediatr.*, 53, 731.
- Potter, J. M. (1955). Angiomatous malformations of the brain: their nature and prognosis. *Ann. roy. Coll. Surg. Engl.*, 16, 227.
- Ray, B. S. (1941). Cerebral arteriovenous aneurysms. *Surg. Gynec. Obstet.*, 73, 615.
- Reid, M. R. (1925a). Studies on abnormal arteriovenous communications, acquired and congenital. Part 1. Report of a series of cases. *Arch. Surg. (Chicago)*, 10, 601.
- (1925b). Abnormal arteriovenous communications, acquired and congenital. Part 3. The effects of abnormal arteriovenous communications on the heart, blood vessels and other structures. *Ibid.*, 11, 25.
- Russell, D. S. and Nevin, S. (1940). Aneurysm of the great vein of Galen causing internal hydrocephalus: Report of two cases. *J. Path. Bact.*, 51, 375.
- Schmitt, O. (1920). Beiträge zur Klinik und chirurgischen Behandlung des Angioma arteriale racemosum, besonders des Kopfes. *Brunns Beitr. klin. Chir.*, 118, 178.
- Shenkin, H. A., Spitz, E. B., Grant, F. C., and Kety, S. S. (1948). Physiologic studies of arteriovenous anomalies of the brain. *J. Neurosurg.*, 5, 165.
- Silverman, B. K., Breckx, T., Craig, J., and Nadas, A. S. (1955). Congestive failure in the newborn caused by cerebral arteriovenous fistula. *Amer. J. Dis. Child.*, 89, 539.
- Steinheil, S. O. (1895). 'Ueber einen Fall von Varix aneurysmaticus im Bereich der Gehirngefäße.' Inaug. Diss. Würzburg. Quoted by Dandy, W. E. (1928).
- Wolfe, H. R. I. and France, N. E. (1949). Arteriovenous aneurysm of the great vein of Galen. *Brit. J. Surg.*, 37, 76.

Addendum

Since this paper has been sent to the press, two additional cases of cerebral arteriovenous fistula associated with heart failure have been reported by Glatt and Rowe (1960).