

with the patient before and after his operation, he seemed perfectly normal, though apathetic, could and did answer all questions intelligently but now has absolutely no memory of anything from about two weeks before his admission to the hospital until the night he wished to urinate six days after his operation. He does not remember leaving home some sixty miles away and coming to hospital, nor can he recall ever having seen me before the sixth day after his operation. His memory of events since that time is perfectly clear. In conclusion, one cannot help but meditate as to just how great was the rôle played by anti-streptococcal medication (amino-sulphamide) in preventing such complications as meningitis or cerebritis.

CONGENITAL RHABDOMYOMA OF THE HEART*

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Baby O.K., a male of 9 pounds 1½ ounces, delivered normally at term, was admitted to the Hospital for Sick Children on November 20, 1936, at the age of 2 days with a history of attacks of cyanosis developing 10 hours after birth. There had been no difficulty in resuscitation at birth and the child cried lustily. The first attack lasted 1 hour and gradually cleared, but subsequent attacks increased in duration and frequency until at 2 days of age the cyanosis was persistent and generalized. The skin was mottled and the lips cyanosed. Respiration was distressed, rapid and grunting. The chest was large, symmetrical and barrel-shaped. The breath sounds were vesicular, with a few râles over both bases. The area of præcordial dullness was markedly enlarged, extending from the right nipple line to the left lateral chest wall, and from the clavicle to the area of liver dullness. Heart sounds were muffled and indistinct but no murmurs were heard. The heart rate was very rapid and no irregularity was noted clinically, although electrocardiographic tracings were not made. The liver was enlarged almost to the level of the umbilicus; the spleen was also enlarged to 2 finger-breadths below the costal margin. The cyanosis steadily progressed in the oxygen tent and the child died on its third day. X-ray showed an abnormally large heart shadow occupying the major part of the thorax. Red blood cells 6,000,000; white blood cells 19,000; hgb. 120 per cent; calcium 9.03 mg.

Autopsy was performed 12 hours after death. The anatomical diagnoses were: (1) congenital rhabdomyoma of heart; (2) patent foramen ovale and ductus arteriosus; (3) partial atelectasis; (4) hydropericardium; (5) hepatomegaly; (6) splenomegaly.

Over one-half of the thorax was occupied by a tremendously enlarged heart and pericardial sac. The pericardial fluid was excessive, amounting to about 15 c.c. The heart was roughly globular in shape measuring 7 cm. in diameter. A few small circumscribed greyish pink nodules, measuring from 0.5 to 1 cm. in diameter, were visible through the pericardium in the apical region of the left ventricle and also in the auricular walls. On opening the heart a thick distorted inter-

ventricular septum was found bulging into both right and left ventricles. In the right ventricle the septal surface was nodular and warty, projecting into the cavity, which was almost obliterated. The septum was incised longitudinally and a large, round, circumscribed tumour mass was exposed, measuring 4.5 cm. in diameter and surrounded by only a narrow rim of compressed myocardium (Fig. 1). The tumour was clearly demarcated from the muscle by a fine capsule. The cut surface was uniform, soft, moist, putty-like, pinkish grey in colour and everted slightly above the cut edge. Besides the large tumour in the septum, multiple smaller nodules were present. Two nodules, each measuring 1 cm. in diameter lay in the left ventricular wall near the apex; another 0.7 cm. in the right ventricular wall near the base and two 0.5 cm. in the right auricle. The small nodules were similar to the large mass on cut surface. The valve leaflets were thin and pliable and were not involved in the tumours. A small foramen ovale still remained and the ductus arteriosus was patent. The great vessels arose in the usual manner and appeared normal.

The lungs were partially atelectatic in their lower lobes; the liver and spleen were enlarged and congested. Small deposits of uric acid crystals lay in the medulla of the kidneys. Other organs appeared normal.

Microscopic examination.—Sections from various tumour nodules all showed a similar histological picture. They were enclosed with a thin fibrous capsule for the most part, but here and there the tumour cells had broken through and lay adjacent to the muscle cells. The tumour cells had a peculiar lacy appearance due to the presence of large irregular vacuoles. Individual cells were extremely large, many of them approximately the size of a fetal glomerulus. A wide variety of bizarre cells of embryonic character constituted the tumour and no uniformity of type cell could be identified. In shape they were found round, oval or spindle-shaped, with nuclei central, eccentric or compressed to the peripheral cell membrane. The nuclei were large or small, pale or hyperchromatic, many with prominent nucleoli, often vesicular. Many cells showed a centrally placed nucleus about which the cytoplasm was arranged in fine radiating acidophilic fibrils extending toward the periphery, forming the spider cells so characteristic of rhabdomyoma. A few cells contained a diffuse, granular, pink-staining cytoplasm suggesting ground glass. Under very high power many of these cells showed the finely granular structure arranged to form vague cross-striations suggesting those of a striated muscle cell. This character was more clearly elicited with phosphotungstic-acid-haematoxylin stain. Much distortion of cells existed due to large clear vacuoles occupying the main part of the cytoplasm and displacing the nucleus. Many large clearly outlined spaces bore no relation to neighbouring cells and were devoid of nuclear or any stainable material. The contents of these vacuoles could not be determined. In paraffin preparations the vacuoles were empty and clearly defined. Frozen sections stained with Sudan III showed no evidence of true fat within the vacuole, but a few very tiny droplets lay within the cytoplasm of the cell. Mitotic figures and multinucleated cells were not found. Small thin-walled blood vessels were present in moderate numbers. A few fine strands of supporting stroma were interspersed between large groups of tumour cells.

COMMENTS AND CONCLUSIONS

Rhabdomyoma of the heart forms a small group of the many deformities which are present at birth. The important factors in relation to myocardial function are the size of the tumour and its location as related to the conducting mechanism of the heart. The invasive quality of the tumour appears to be practically

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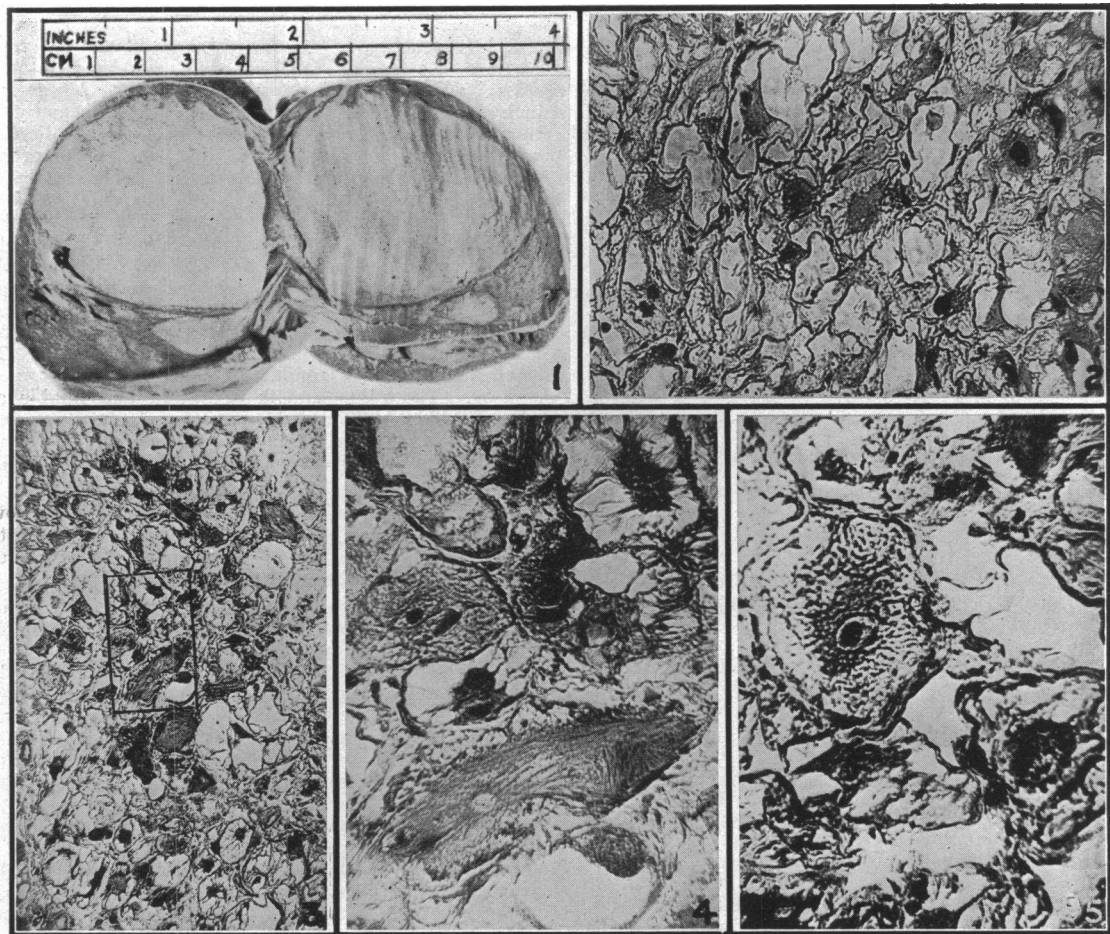


Fig. 1.—Large rhabdomyoma occupying interventricular septum with smaller tumours nearby. Fig. 2.—Lacy appearance of tumour cells. (P.T.A. x 150). Fig. 3.—Cellular structure of rhabdomyoma. (P.T.A. stain x 100). Fig. 4.—Tumour cell showing longitudinal and cross striations. Other vacuolated cells. (P.T.A. x 600). Fig. 5.—“Spider cell”. (P.T.A. x 600).

nil. Pressure and distortion however, interfering with contraction of the muscle fibres as well as conduction of impulses are of greater moment. Although the tumours are not considered genetically related to the Purkinje cells, but rather to the pure muscle cells (Hueper), it is unlikely that conduction could remain undisturbed in proximity to a tumour of such dimensions as the above.

This case of rhabdomyoma adds one more to the relatively small number in the literature (49). The salient features of the case are the multiplicity of tumours in the myocardium (6),

the remarkable size of the mass in the septum, the associated large liver and spleen, and the embryonic type of cell with the lack of malignant histological characters.

BIBLIOGRAPHY

1. WOLBACH, S. B.: Congenital rhabdomyoma of heart, *J. Med. Res.*, 1907, 16: 495.
2. FARBER, S.: Congenital rhabdomyoma of the heart, *Am. J. Path.*, 1931, 7: 105.
3. BERGER, L. AND VALEE, A.: Congenital rhabdomyoma of heart, *Annal. d'Anat. Path.*, 1930, 7: 797.
4. WEGMAN, M. F. AND EGBERT, D. S.: Congenital rhabdomyoma of heart associated with arrhythmia, *J. Pediat.*, 1935, 6: 818.
5. HUEPER, W. C.: Rhabdomyomatosis of the heart in a negro, *Arch. of Path.*, 1935, 19: 372.
6. PAULLI, W.: Congenital diffuse rhabdomyomatosis of heart in two brothers, *Monatschr. f. Kinderh.*, 1936, 66: 29.
7. TAMURA, O.: Rhabdomyoma of heart, *Gann.*, 1936, 30: 391.

OPHTHALMIC INDICATIONS FOR ABORTION. — A. Favory is of the opinion that pregnancy should always be terminated in the presence of retinitis of renal origin; not only the possibility of blindness but also the condition of the kidneys are the deciding factors. In retrobulbar neuritis, syphilis and nasopharyngeal conditions having been eliminated, induction should be considered. Myopia is often aggravated by pregnancy, and in these cases retinal hæmorrhages and

detachment sometimes occur. Pregnancy should be forbidden where there is already detachment in one eye. All forms of choroiditis, uveitis, and keratoconus are made worse; in the first of these abortion should be considered. In hereditary types of eye disease which may involve blindness the necessity for intervention is not yet generally admitted. Here the eugenic and legal aspects of the case must influence the decision.—*Progr. Méd.*, Paris, January 29, 1938, p. 153. Abs. in *Brit. M. J.*