Congenital contraction of orifice of pulmonary artery from fusion of the valves : foramen ovale open / by Judson S. Bury.

CONGENITAL CONTRACTION

OF

ORIFICE OF PULMONARY ARTERY

FROM FUSION OF THE VALVES:

FORAMEN OVALE OPEN.

Sequel to "A Case of Extreme Cyanosis in an Adult."

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SEQUEL TO

"A CASE OF EXTREME CYANOSIS IN AN ADULT."[1]

A CYANOTIC woman, aged twenty-one years, was exhibited at the Manchester Medical Society, Oct. 4th, 1882; and details of her condition are given in THE LANCET of the date mentioned, of which the following are the more important:—

Mary F—— had good health till between eighteen and nineteen, when, after hurrying a good deal, she had pains in the chest and back and a choking sensation, and was told that "she was as blue as a whimberry." After a temporary disappearance or abatement of the blueness it returned and persisted. The chest wall was unduly prominent opposite the second, third, and fourth left rib cartilages. "The jugular veins are not distended, but there is distinct pulsation of the carotid arteries. The heart's impulse is diffused and heaving; it is felt in the third, fourth, and fifth spaces, as far out as the anterior border of the left axilla, but is not felt near the sternum, nor is there

[1] Reported in THE LANCET, 1882, vol. ii., p. 801.

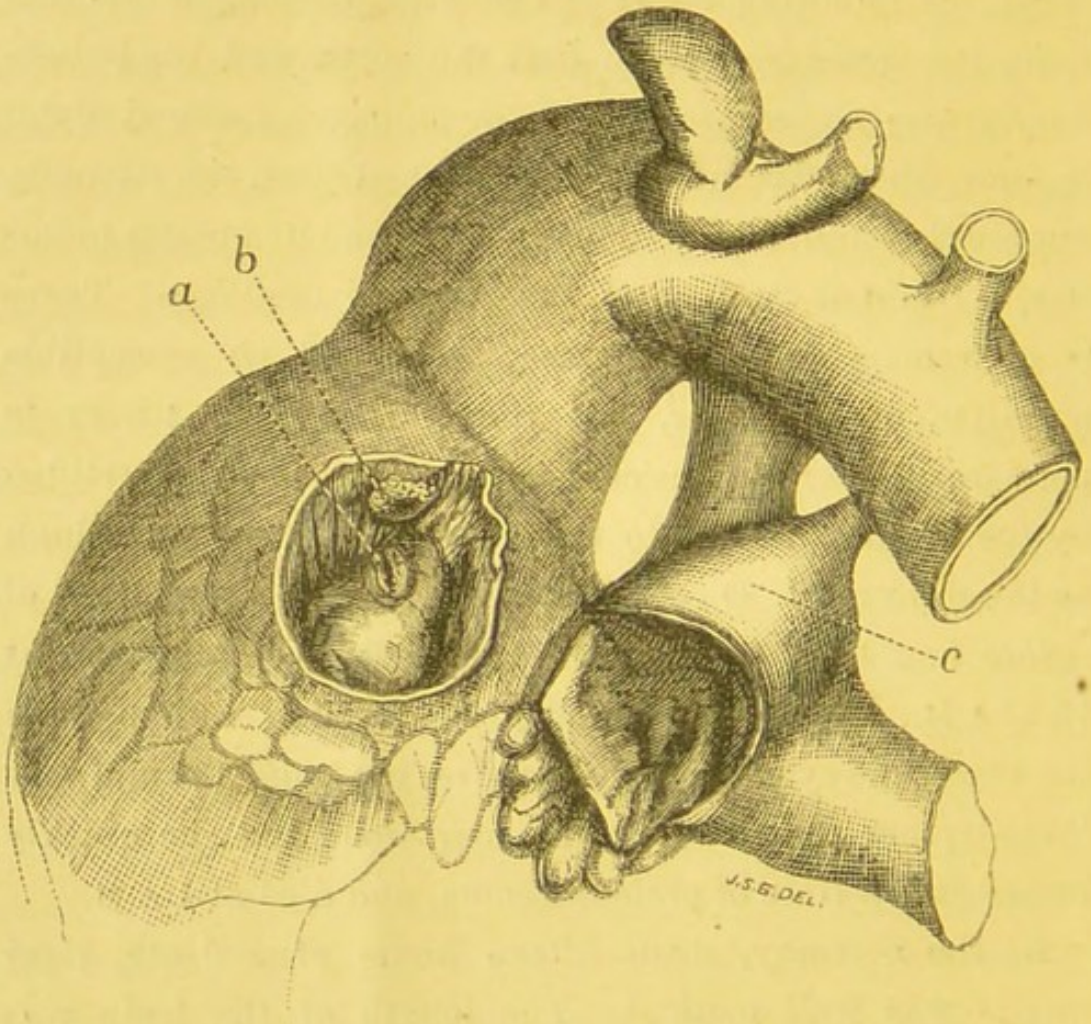
any epigastric pulsation. The maximum point of the impulse is in the fifth space in the nipple-line; no thrill. The cardiac dulness is much increased, and laterally rather than vertically, extending from about one inch to the right of the sternum to almost the middle of the left axilla; the third rib marks the upper limit of superficial dulness, though the first two spaces are higher pitched than those on the right side; the sixth rib is the lower limit. On auscultation a high-pitched, systolic, whistling murmur is heard at the base, of maximum intensity over the first piece of the sternum; it is well conducted down this bone, being also well heard at the right sterno-clavicular articulation, and faintly in the carotids. A vertical line from the middle of the left clavicle marks its extreme left limit, and the murmur is gradually lost as the stethoscope is moved towards the apex; it is not heard behind; the sounds at the left apex are quite pure. The second sound is accentuated over the pulmonary cartilage, and is louder than at the aortic cartilage. The pulse is regular in rhythm, but not quite regular in force; it is equal on the two sides, and is felt in the femorals and in other arteries."

The patient was seen again in February, 1883, when she stated that she had had attacks of fainting for about a month, and that the pain in her chest was worse; it was noted that the cyanosis had increased, and that the systolic basic murmur was not so loud as formerly, but the heart in other respects was unaltered. The chest pains and difficulty in breathing became more severe, and early in April she was admitted into the hospital. On April 5th, at 1 A.M., the house-surgeon, Mr. Buchan, was called to her: "she was insensible, breathing stertorously, the pulse was rapid and very thready; there was no rigidity or spasm of muscles. Mus-

tard poultices were applied to the cardiac region and an enema of brandy administered. The patient remained unconscious for half an hour and then slowly recovered, answered questions, but was very drowsy and heavy for some time. Temperature 100·8°. Later on in the day she seemed much as usual, and did not complain of anything." On April 23rd my colleague, Dr. Railton, carefully examined the heart and made the following note: "The basic systolic murmur is faint, it is heard equally in both the aortic and the pulmonary areas; no accentuation of the pulmonary second sound is perceptible. The murmur is continued down the sternum, but another murmur, systolic in time and of greater intensity, is heard at the base of the ensiform cartilage. There is no venous pulsation in the neck, and no perceptible pulsation of the liver, but the liver is very tender to pressure. The right auricle is largely distended, as testified by the absolute dulness to the right of the sternum as high as the third rib." On April 30th the patient complained of severe headache, especially at the back. On May 1st, at 12.20 A M., Mr. Buchan found her screaming with pain in the chest, the cyanosis was excessive, the pulse 130 and very thready, her breathing became stertorous and she quickly passed into a state of profound coma, and died at 1 A.M.

At the necropsy, made fifteen hours after death, rigor mortis was well marked. The length of the body was about fifty-six inches, the limbs were well formed and the subcutaneous fat was everywhere abundant. The lips, ends of fingers, and toes were of a dark-purple colour. On removing the sternum and costal cartilages the pericardium came prominently into view, the tip of the lower lobe of the right lung was also seen, but the left lung was out of sight, being shrunk and bound down by firm fibrous bands. Trans-

versely the exposed pericardium measured six inches and a half, and vertically in the mid-sternal line nearly four inches. The liver was much enlarged, had its lower margin in the same line four inches and a half below the pericardium. On opening the latter the heart, of large size, was placed with its longest diameter transversely, it was also tilted; its



anterior surface was made up of right auricle and ventricle, the left ventricle being out of sight. The outer margin of the right auricle was two inches to the right of the right edge of the sternum; both the right cavities were distended with black blood. The heart separated and emptied weighed seventeen ounces, just double the average weight

in females at this age; it was square in shape and measured five inches from the root of the pulmonary artery to the apex, and four inches and a half from side to side. The four pulmonary veins were traced to the left auricle. The pulmonary artery after dividing normally into two branches appeared to be prolonged by means of the ductus arteriosus into the aorta; but subsequent dissection showed that there was no opening into the latter, but that the lower part of the duct consisted of a conically shaped space (see figure, *c*), the closed apex being three millimetres or one-ninth of an inch from the aorta. The valves of the pulmonary artery were blended, and formed a thick dome-shaped projection into the root of the artery, dividing the space at its base into three deep pockets. At the top of the dome was a slit-like aperture (*a*), one-eighth of an inch in length, and admitting easily the end of an ordinary probe; it was lined by minute vegetations, and on the wall of the artery immediately above and on the aortic side of this aperture was a small raised cluster of vegetations (*b*). The opening mentioned formed the only direct communication between the right ventricle and the pulmonary artery; the latter and its branches were smaller, and had thinner walls than normal. The right auricle and ventricle were dilated and hypertrophied, the muscular substance of the ventricle being dark-red and unusually firm and thick, and the musculi papillares very large. The tricuspid orifice admitted the tips of two fingers; granulations covered with fibrin were found on two of the segments of the valve: namely, on the left one—that resting on the ventricular septum—and on the anterior one, in each case on the auricular surface and near the lower edge, the crop of vegetations on the anterior cusp being

much the larger and a quarter of an inch in diameter. The coronary sinus appeared much larger than natural; its auricular end was nearly half an inch in diameter. The foramen ovale, widely open, measured seven-eighths of an inch vertically and five-eighths across, a portion of the valve with a sharp edge projected from the posterior and lower side of the foramen. The left were, as compared with the right cavities, small in capacity; the ventricular wall was thickened, and its maximum thickness was a little greater than that of the wall of the right ventricle (for measurements see below). The left auriculo-ventricular opening was not quite so wide as the right one, scarcely admitting the tips of two fingers; both segments of the mitral valve were thickened just above their free edges, but there were no signs of recent endocarditis. The aortic valve presented slight irregularities; the two posterior flaps were firmly united for a short distance from their common angle of origin; they were also fenestrated near their junction with one another and with the anterior flap, at each corner of each of these valves were two small holes crossed by a slender fibrous band; the lunulæ of the anterior cusp were entire. Above the valve small patches of atheroma were present on the wall of the artery. Both the aorta and pulmonary artery were unusually small, alike as regards their calibre and the thickness of their walls; the aorta looked slightly constricted after giving off the left subclavian, and the lumen here was half an inch less than at the origin of the aorta. There were two instead of three primary branches, the left carotid and innominate starting as a common trunk. The accompanying table of measurements brings out, amongst other things, the contrast between the size of the heart and that of its orifices and the two arterial trunks; the former is

larger, and its walls much thicker, the latter smaller and thinner, than natural.

*Dimensions of the heart, in inches.**

	Healthy female.		Mary F——.
Circumference of heart	9·236	....	11·125
Girth of right ventricle	5·184	....	7·125
Girth of left ventricle	4·049	....	4·
Thickness of walls of right ventricle	·118—·164	....	·4—·5
Thickness of walls of left ventricle ..	·222—·497	....	·4—·62
Thickness of septum	·421	....	·5
Circumf. of right auriculo-ventricular aperture	4·562	....	3·5
Circumf. of left auriculo-ventricular aperture	3·996	....	3·
Circumf. of pulmonic aperture	3·493	....	2·5
Circumf. of aortic aperture	3·019	....	2·
Circumf. of lumen of descending portion of arch	—	....	1·5

Weights.

	Healthy female.		Mary F——.
Heart	8¾ ounces	....	17 ounces
Lungs	32 „	....	30 „
Kidneys	8—9 „	....	15 „
Spleen	4—9 „	....	8 „
Liver	40—50 „	....	54 „
Brain	43·7 „	....	49½ „

* See Peacock's article in Reynolds' System of Medicine, vol. iv.

The lungs, much congested, were free from tubercle. The kidneys, spleen, liver, and brain were also very congested, and, as seen in the table, heavier than natural. Slices of the brain exhibited black bleeding points. The liver, large and firm, presented the characteristic nutmeg appearance, and showed under the microscope great dilatation of the veins, especially of the intralobular ones, and around the latter the liver cells were much wasted, the intervening spaces being filled up with yellowish-brown pigment

granules. There was also a small-celled infiltration of the periportal connective tissue. The kidneys were firmer than natural, and sections showed a commencing intertubular cirrhosis, and all the bloodvessels and large tracts of tubules were filled and distended with red blood-corpuscles.

Remarks.—The view taken of the case during life—namely, "that the origin of the pulmonary artery was affected with endocarditis in fœtal life; that the effects of its obstruction were compensated for by the persistence of communication between the two sides of the heart, and perhaps also by a patent ductus arteriosus"—is shown by the post-mortem examination to have been in the main correct. But now, further, we see, from the absence of any defect in the septum between the ventricles, that in all probability the pulmonary artery became diseased late in fœtal life, after the completion of the septum, and that at birth the contracted orifice rendered the artery incapable of transmitting the increased current of blood to the lungs, and then the ensuing strain on the right ventricle would keep open and be relieved by the foramen ovale. With so much obstruction at the exit of the right ventricle the relatively large size of the pulmonary artery and its branches appears somewhat remarkable, and one would expect to find a patent ductus arteriosus or some other channel by means of which blood could enter the artery. But perhaps the narrowing of the pulmonic aperture from blending of the semilunar cusps was slow enough to permit the closure of the duct, and attained its maximum some time after this closure; and even with a small communication between the ventricle and artery, the latter would be kept full of blood, and so maintain a fair size or dwindle very slowly. In the previous report it was pointed out that many of the physical signs suggested obstructive disease at

the aortic orifice, thus the systolic murmur loudest at the top of the sternum was traceable towards the inner end of the right clavicle, and along the carotids; there was no epigastric pulsation, but a heaving impulse in the left mammary region. The autopsy revealed excessive hypertrophy with some dilatation of the right ventricle, and moderate hypertrophy of the left ventricle associated with a partial fusing of two cusps of the aortic valve; the frænum thus formed projecting into the aortic aperture would be likely to evolve sonorous vibrations in the blood of the ascending arch, and at least may be offered as an explanation of the fact that a systolic bruit was heard as well below the right as below the left clavicle, in the latter position generated with greater certainty by the narrowed pulmonic orifice. In the presence of such extreme right hypertrophy the absence of any perceptible transmitted impulse to the lower end of the sternum or to the epigastrium, is a striking clinical feature, and one to be remembered in making a diagnosis of unusual heart conditions. The chest was repeatedly examined, and yet no pulsation was ever felt near the sternum, and the maximum point of the heaving impulse was always about the left nipple-line. These signs receive some interpretation by a consideration of the rotated transverse position of the heart with its concealed hypertrophied left ventricle. The fenestration of the aortic valve is probably a congenital defect, and would not give rise to much, if any, regurgitation. The small size of the arch is noteworthy, and especially the constriction of the third portion. During the last few weeks of life a systolic murmur was heard at the ensiform cartilage, and the pulmonary second sound became weaker. The fibrinous deposits on the tricuspid valve and the result-

ing imperfect adaptation of its segments give reasons for a tricuspid murmur; and it has been said that tricuspid regurgitation will lessen the intensity of the second sound at the pulmonary cartilage.[1] Probably at this time, too, endocarditis was lit up in the pulmonary artery. Such gradually increasing obstacles to an already over-strained heart fully account for the increasing distress, the final paroxysms of pain, the quickly ensuing comatose death, and for the intense venous congestion of the brain and other organs found after death.

Many similar cases to the one related are to be found in medical literature. Thus, Dr. Peacock[2] collected notes of twenty persons who presented this anomaly — namely, obstruction at the orifice of the pulmonary artery with an open foramen ovale, and eleven of these lived to the age of fifteen years and upwards.

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1 See Balfour: Clinical Lectures on Diseases of the Heart and Aorta, second edition, p. 194.

2 Malformations of the Human Heart, second edition, p. 180.