Survival to the Age of Seventy-Five Years with Congenital Pulmonary Stenosis and Patent Foramen Ovale

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It is very rare for a person with cyanosis and finger clubbing due to congenital heart disease to reach old age. Survival beyond the age of 50 has been noted only a few times and, as far as we are aware, survival of a person with morbus caeruleus beyond 70 years is unique. The patient reported below showed at autopsy marked pulmonary stenosis and patency of the atrial septum simulating the tetralogy of Fallot. One of the reasons for the long survival was undoubtedly a markedly developed collateral bronchial circulation.

The syndrome of pulmonary stenosis with patency of the foramen ovale is a distinct entity though readily confused clinically with the tetralogy of Fallot. The oldest previously recorded case proved at autopsy was 57 years old. It is therefore of particular interest to report here with the case of a woman with this congenital anomaly who not only survived to the age of 75 years but who also lived a useful and comfortable life despite considerable cyanosis until death from cardiac and cerebral vascular failure.

Pulmonary stenosis with patency of the foramen ovale was one of the first congenital anomalies to be recognized. Morgagni described a girl aged 16 years with this condition in 1761. Six more cases were reported during the next 100 years. We have recorded chronologically in the bibliography more cases since Wilk's paper of 1859. Bertin and Breschet's case, quoted by Gintrac and Lallemand, the oldest previously recorded, has been referred to several times as though it were new each time. Among 37 of this group of 38 cases the ratio as to sexes was 16 males to 21 females and the ages at death were in the first decade in 9 cases, in the second in 16, in the third in 8, in the fourth in 2, in the fifth in 1, and in the sixth in 1. Our case died in the eighth decade.

We would like to emphasize that pure pulmonary stenosis, pulmonary stenosis with patency of the foramen ovale, and pulmonary stenosis with ventricular septal defect and dextroposition of the aorta (tetralogy of Fallot) are three distinct clinical entities with different clinical and laboratory findings. In addition, the prognosis is not the same for all three and the surgical approach in the three conditions also differs. It is, therefore, important to differentiate and to report all three syndromes as separate conditions and no longer to group them under general "pulmonary stenosis."

The clinical features of this syndrome have recently been reviewed by Selzer and his associates: Cyanosis, clubbing of the fingers, polycythemia, loud systolic murmur at the pulmonary area, right ventricular hypertrophy, prominent P waves in the electrocardiogram, enlarged x-ray shadow of the pulmonary artery with moderate-sized hilar shadow. These findings differ from the tetralogy of Fallot with respect in particular to the x-ray evidence; the typical x-ray heart and vascular shadow in the tetralogy of Fallot shows a small pulmonary artery and small hilus shadows. It differs from pulmonary stenosis with closed septa with respect to the clinical findings of cyanosis, clubbing, and polycythemia, which are absent in the latter except for terminal cyanosis when heart failure develops.

Case Report

M. T. M., born May 29, 1874, was first examined by one of us (P. D. W.) on April 24, 1922. At that time (at the age of 48) she gave the following history.

Heart disease had been known since early childhood but the patient did not think that she had been blue at birth. Cyanosis gradually developed in her early youth and was present in considerable degree later. Shortness of breath was an early symptom and limited her activity in playing with

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Circulation, Volume II, October 1950
other children. Her heavy breathing always revealed her hiding place to the others in their game of hide and seek. This shortness of breath on effort continued to be a constant symptom throughout her life. She had recently had her teeth extracted, two at a time, without complications.

Her general health had always been rather frail, and she had had pneumonia at the age of 18, typhoid fever at 19, severe “jaundice” at 21, and influenza at 33. The last named illness was followed by marked "nervous exhaustion." At that time, 1907, because of fear of heart failure she was digitalized for the first time and had continued to take digitalis ever since. There had been no chest pain, hemoptysis, or ankle edema.

She had worked steadily for many years as a librarian and secretary. For six years (1912–1918) she had assisted in the record room at the Peter Bent Brigham Hospital. While there she was at one time examined by Dr. Maude Abbott who diagnosed deviation of the aorta and possible pulmonary stenosis.

Family history revealed that she was an only child. Her father died at the age of 75. Her mother was 76 years old at this time (in 1922) and in good health; she lived to be 88 years old.

Physical examination showed a rather short, otherwise well developed and nourished middle-aged woman of 48, alert, breathing normally with moderate cyanosis of the face, lips, tongue, and finger tips. There was doubtful clubbing of the fingers. The pulse was regular at 80. The blood pressure was 110/85. There was no precordial thrill palpable. The cardiac apex impulse was faint in the fifth intercostal space, 0.5 cm. outside the left midclavicular line. The first sound was fairly loud at the apex, but the second sound was diminished throughout. There was a loud blowing systolic murmur heard all over the precordium, loudest in the second left intercostal space near the sternum. There were no diastolic murmur. The lungs, abdomen and the remainder of the physical examination were negative. There was no edema of the extremities.

For the electrocardiogram see figure 1A.

A diagnosis was made of congenital pulmonary stenosis without heart failure. Since there was no evidence of congestive failure, digitalis was discontinued.

The next examination was on September 26, 1931. She had been quite well during the intervening nine and one-half years, still working as a librarian and secretary, and although she was bothered by shortness of breath and occasional palpitation, her health was fairly good.

Physical examination revealed that the heart had increased slightly in size. The murmur in the pulmonary area was of about grade 4 intensity and on this examination, a slight systolic thrill could be felt in the pulmonary area. The cyanosis of the face and finger tips was still evident and on this examination slight clubbing of the fingers was definitely noted.

Laboratory Data (Sept. 26, 1931): The red blood count was 6,750,000. The hemoglobin was 90 per cent (Sahli). The white blood count was 7,850.

Fluoroscopic examination showed slight to moderate cardiac enlargement. The transverse diameter of the heart was 13.4 cm. and the internal diameter of the chest was 22.1 cm. The distance across the great vessels at the base of the heart was 6.5 cm. The hilar vascular shadows were well seen.

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**Fig. 1.** A. 4/24/22. Electrocardiogram showing a rate of 80, marked right axis deviation, prominent P waves, and inverted T1 and T4. B. 9/26/31. Electrocardiogram showing a rate of 75, marked right axis deviation, diphasic T1 and inverted T4. The voltage is less than on 4/24/22.

For the electrocardiogram see figure 1B.

In 1931 an adult cyanotic patient with the findings as listed above was considered to have the tetralogy of Fallot, and therefore this diagnosis was made on this examination.

She continued to be fairly well from 1931 until 1949. The physical examination changed very little except that the cyanosis and finger clubbing increased gradually through the years.

On March 19, 1949, she was seen by Dr. Charles Walcott of Cambridge, Massachusetts, because of her complaints of increasing dyspnea and edema of the legs. Right heart failure was diagnosed and
digitalis, low-sodium diet, and rest were prescribed. After a few days she improved.

From April 3, 1949, until May 7, 1949, she had several episodes of dyspnea and syncope. During this period the pulmonic systolic murmur decreased greatly in intensity.

On May 6, 1949, she had a paroxysm of auricular fibrillation with a ventricular rate of 96 for which 0.2 Gm. of quinidine sulfate were given. The paroxysm stopped within 20 minutes. The syncopal attacks continued until she died suddenly at 12 noon on May 7, 1949. The cause of death was considered to be heart failure associated with cerebral ischemia and possibly cardiac arrhythmia.

Autopsy. The autopsy was performed eight hours after death by one of us (R. H. F.).

The body was that of an elderly woman measuring 150 cm. in length and weighing an estimated 130 pounds. The entire body was cyanotic with marked lividity of the dependent portions. There was minimal clubbing of the fingers. Slight pitting edema of the ankles was present.

The pericardium contained a few cubic centimeters of amber fluid. The heart weighed 390 grams. The right ventricle was almost as large as the left. The right atrium was three times normal size and its wall varied from 2 to 3 mm. in thickness. The left atrium did not appear enlarged. The foramen ovale was patent, the defect measuring 1.2 cm. in greatest diameter. The guard on the left side covered approximately one-third of the defect. At the edges of the opening the endocardium was thickened and gray. On the left side a fibrous cord 1 mm. in diameter extended across the foramen anteroposteriorly and was attached near the edges. The right ventricular wall averaged 12 mm. in thickness, but varied from 18 mm. in the conus to 5 mm. near the apex. The columnae carneae were prominent. The myocardium of the left ventricle was 18 mm. thick near the base but tapered to 10 mm. at the apex. The distance from the atrioventricular valve ring to the apex of the chamber was 5 cm. on the right and 6.5 cm. on the left. The myocardium was red and firm except in the anterior wall of the right ventricle where the wall was thinned and there were prominent gray strands of tissue, especially conspicuous beneath the endocardium. The endocardium of the right ventricle was gray and opaque and this became very marked near the pulmonic ring.

The infundibulum of the right ventricle narrowed gradually as it approached the pulmonic ring. The narrowing was accentuated by the markedly hypertrophied columnae carneae and just proximal to the pulmonic ring for a distance of 5 mm. the maximum diameter of the infundibulum was 8 mm. A true subpulmonic stenosis was thus manifest. The pulmonic ring was 10 mm. in diameter and the artery just distal to the ring was 20 mm. in diameter. Putting into the lumen of the base of the pulmonary artery the markedly stenotic pulmonary valve which was a dome-shaped structure and measured 20 mm. in length. At the apex of this structure was the valve orifice which was 3 mm. in diameter (fig. 2). About the rim of the opening were adherent, tiny, grayish-pink vegetations. The valvular structure was tough, fibrous, uniformly smooth, and 1 mm. thick throughout. There were 3 attachments of this structure to the wall of the pulmonary artery equidistantly placed around the circumference of the artery, and these were interpreted as probably representing valve commissures (fig. 2). The right posterior attachment extended to within 4 mm. of the opening, the other two to within 8 mm. Between these attachments were recesses extending down to the valve ring between the wall of the artery and the dome-shaped projection.

The chordae tendineae of the tricuspid valve were slightly thickened. Near the middle of the anterior cusp was an area of calcification 3 mm. in diameter which protruded on the atrial surface. The tricuspid valve ring was 9.5 cm. in circumference, the mitral 8.5 cm., and the aortic 7.5 cm. The latter two valves were normal in appearance.

There was minimal atherosclerosis of the coronary arteries but no evidence of an old or recanalized thrombus was seen.

The aorta was normal in size and position and showed moderate atherosclerosis. The pulmonary artery was 2 cm. in diameter just above the pulmonic valve ring but dilated gradually so that the primary trunk at the bifurcation was 5 cm. in diameter. The dilatation ended abruptly at the bifurcation.

The lungs were well aerated and the pleural
surface was a variegated blue-red. There were numerous fibrous adhesions to the chest wall. The trachea contained a profuse amount of pink, frothy fluid. The bronchial arteries were injected in situ via the aorta after ligation of its main branches. A colored radiopaque plastic solution in acetone was used.*

Later the pulmonary arteries and entire bronchial tree were injected (fig. 3). X-ray films were taken after each injection and the lungs were finally digested in hydrochloric acid. The bronchial arteries arose from the aorta, both at the arch and from the descending aorta and anastomosed around the hilar region to form a network of vessels which proceeded out to the lung parenchyma, most of them in direct relationship to the bronchi. Some of these vessels measured 6 to 8 mm. in diameter. As they coursed along the bronchi, anastomoses with the pulmonary artery were seen in at least 7 places. The continuity of the two circulations was definitely established by the mixing of the two colored injected masses. A bronchial artery 3 mm. in diameter arose from the plexus of esophageal and bronchial vessels and

Fig. 3. Photograph of injection of pulmonary arteries and entire bronchial tree. Posterior view of digested cast. Note large number of bronchial arteries entering right lung from descending aorta. Also note the huge pulmonary artery to the left of the aorta exposed by clearing away some of the lung and vessels overlying it.

* The authors wish to express their gratitude to Drs. B. H. Landing and James T. Weston of the Childrens Hospital, Boston, for the injection and examination of the lungs.
passed directly into the pulmonary artery bulb posteriorly. In several areas bronchial vessels left the bronchi to course into lung parenchyma and form a small bulb or plexus of vessels far out in the periphery with no precapillary anastomoses to pulmonary vessels. Small branches of the pulmonary artery continued outside of the lung in the adhesions. In the lower portions of the lung there was a definite, somewhat generalized tubular dilatation of the smaller bronchi and bronchioles.

The liver weighed 850 Gm. and the surface was ragged with fibrous tags. It showed increased resistance but the architecture was normal. The gastrointestinal tract, spleen, pancreas, adrenals and kidneys were not unusual. The left wall of the uterus was partially replaced by soft, reddish-gray, granular tissue but the cervix was not involved. The tubes and ovaries were normal.

Microscopically the myocardium of the right ventricle was extensively replaced by fibrous tissue, the inner half of the wall being most severely damaged. In the thinned area on the anterior wall only scattered muscle fibers remained and the wall consisted almost entirely of dense scar tissue. The muscle fibers that remained varied in size and shape and on cross section the nuclei were bizarre in shape. The vessel walls were thickened, some eccentrically, and primarily by intimal proliferation. Medial thickening and scarring and reduplication of the internal elastic lamina were evident in the arteries. The endocardium was thick throughout the right ventricle but this was especially marked along the anterior wall and about the apex where there were thrombi of varying ages in the crevices between the columnae carneae. The older ones were almost completely organized and consisted of loose connective tissue, and recanalization about the columnae had taken place. In the left ventricle the changes were minimal. Only a few patches of fibrous tissue were seen in the myocardium.

The pulmonic valve was also examined microscopically. It was thickened and consisted of dense collagen with areas of myxomatous degeneration. The vegetations consisted of hyalinized connective tissue with adherent clumps of fibrin. No bacteria were seen. Small calcium deposits were in the valve near the tip. The tricuspid valve also contained myxomatous areas and was thickened.

The lungs showed the large alveoli and the club-shaped ends of ruptured septa characteristic of emphysema. Several areas of fatty replacement suggested healed focal pneumonitis. There were no significant changes in the vessels.

The liver was normal except for increased numbers of round cells in the portal areas. There was no evidence of chronic passive congestion. The kidneys showed mild nephrosclerotic changes. The wall of the uterus was invaded by a well-differentiated adenocarcinoma.

Discussion

The record for longevity established by this case was extraordinary in view of the very high degree of valvular and infundibular pulmonary stenosis associated with a patent foramen ovale. Familial longevity was in her favor, as was the fact that she lived a careful life and was never under very severe strain. Also the evolution of her signs suggested that her pulmonary stenosis may have slowly increased in degree in the course of years. Finally, the compensation received through the bronchial circulation, as found by the injection of the blood vessels of the lungs, was undoubtedly an important factor in her survival.

It is of great interest that she was alert and mentally keen through practically all her life. She was an unusually bright and witty person and even within a few months of her death still showed her mental acuity in conversation. Thus, despite such a handicap, she made herself a useful citizen as well as a long-lived one, much as had Henry Gilbert, described by White and Sprague in 1923 who, with the tetralogy of Fallot, survived to the age of 59 years and 8 months.

Finally, it is noteworthy that she had no actual heart failure until a few months before her death and that her cerebral circulation was maintained satisfactorily until a few weeks before she died when she began to have syncopal attacks.

Summary

We have reported herein the case of a woman with congenital pulmonary stenosis with a patent foramen ovale who survived to the age of 75 and who lived a useful life, death finally occurring a few months after the beginning of failure of the right ventricle associated with insufficiency of the cerebral circulation. The oldest case proved at autopsy previously recorded died at the age of 57.

Appendix

Of 8 cases of pulmonary stenosis recently reported by Allanby and Campbell,7 of which were autopsied, 4 showed patency of the foramen ovale in addition. The oldest case was a woman aged 29 at death.
ACKNOWLEDGEMENT

The authors take pleasure in thanking Dr. R. S. Schaf for his aid in looking up some of the historical references to this condition.

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_Circulation_. 1950;2:558-564
doi: 10.1161/01.CIR.2.4.558

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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