# A CASE OF HEMITRUNCUS ASSOCIATED WITH THE TETRALOGY OF FALLOT

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Hemitruncus is a condition in which one pulmonary artery arises from the right ventricle and the other lung is supplied by a vessel which originates directly from the aorta as in *truncus arteriosus*. Our case has this pattern, but has also an infundibular pulmonary stenosis, a large ventricular septal defect and an overriding of the aorta (Tetralogy of Fallot).

## *Case Report*

F. B. (65/26129), a four year old girl, was brought to our clinic on June 12, 1965 with complaints of tiring easily and cyanosis. She was the product of an uneventful pregnancy followed by a normal spontaneous delivery. No cyanosis was noted at birth nor during the following years. Her physical development was fairly normal; she had walked at about one and a half years of age. About one year previous to admission the parents noticed that she tired easily, showed shortness of breath and some degree of cyanosis on exertion. Her family history was not contributory.

**Physical Examination :** Weight was 15 kg. height, 92 cm., pulse 120/min. Her blood pressure was 100/65 mm Hg, Hemoglobin, 21.30 gm. 100 ml. Lips, fingertips and tongue were definitely cyanotic and a degree of clubbing was present. However she did not look acutely ill. No thrills were felt. On auscultation, the heart sounds were normal and no murmurs were audible. Study of x-ray with barium swallow showed the heart to be normal in size, the pulmonary conus was absent and the pulmonary vasculature was somewhat decreased. The electrocardiogram revealed right axis deviation, right ventricular hypertrophy and right atrial enlargement (Figure 1).

Cardiac catheterization and angiocardiography were performed on August 11, 1965 using the right saphenous vein. The right atrium, left atrium, right ventricle and left ventricle were entered, but the pulmonary artery could not be reached. Catheterization data are shown in the following table.

As seen in the table, there was a right to left shunt at the ventricular level, pressures in both ventricles were equal and a considerable arterial unsaturation was present. Angiocardiography was

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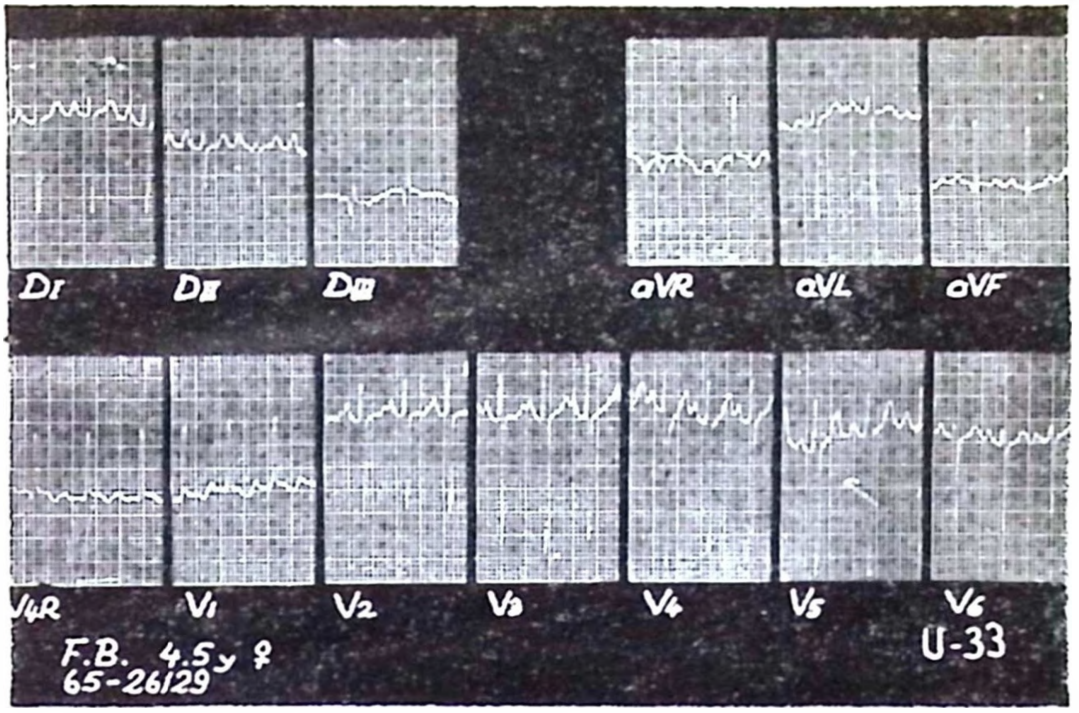


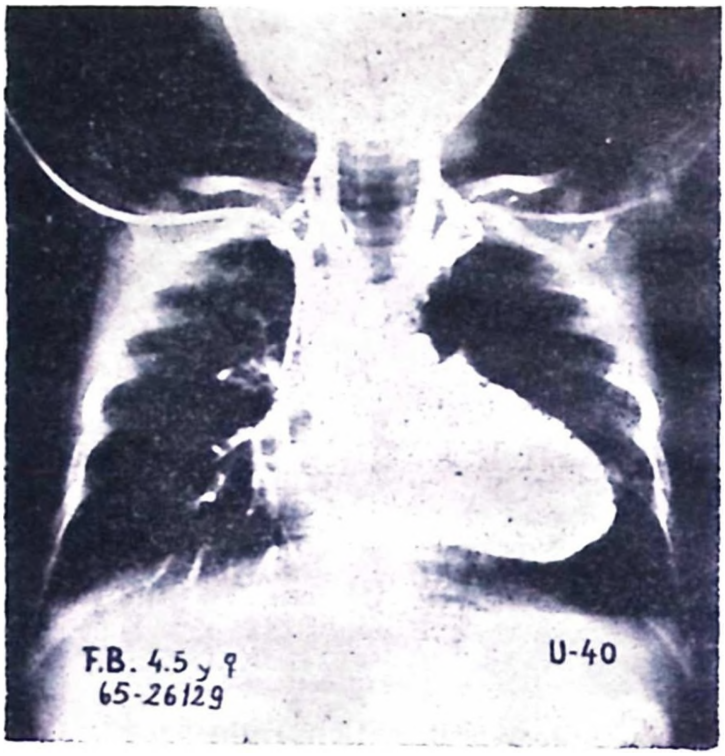

**Figure 1.** Reveals right axis deviation, right ventricular hypertrophy and right atrial enlargement.

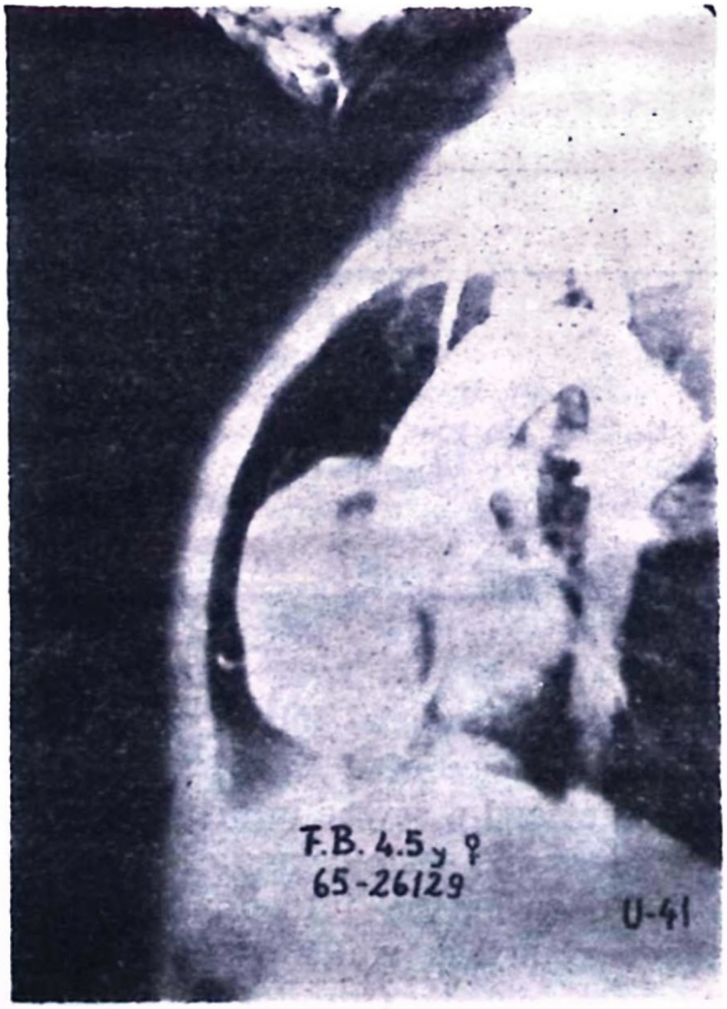
## TABLE 1

PATIENT'S CATHETERIZATION DATA

| | $0_2$ % saturation | pressure (mm Hg) |
|---|---|---|
| S.V.C. | 47 | — |
| Right atrium | 49 | — |
| Left atrium | 94 | 6 (mean) |
| Pulmonary vein | 98 | — |
| Right ventricle | 54 | 102/6 |
| Left ventricle | 85 | 102/6 |
| Femoral artery | 60 | — |

done by injecting opaque material into the right ventricle. Seriograms disclosed the presence of a large ventricular septal defect with right to left shunt and a severe infundibular pulmonary stenosis. The aorta was large and some degree of overriding was present (Figures 2 and 3). Following the infundibular pulmonary stenosis was a single pulmonary artery, which supplied only the right lung. A separate large vessel, arising from the proximal part of the descending aorta branched throughout the entire left lung.




**Figures 2 and 3.** X-rays of the aorta showing a degree of overriding.

*Discussion*

The absence of one of the pulmonary arteries is a well known congenital anomaly in which the involved side is usually supplied by bronchial arteries. In the first case, reported by Fraentzel in 1868, the absence of the right pulmonary artery was associated with aortico-pulmonary window. Following this more than 40 cases were reported. It was more commonly noted on the right side.[2] The absence of the right pulmonary artery shows different features from the absence of the left. When the right pulmonary artery is absent, the right lung is supplied by either bronchial arteries or a single vessel arising from the ascending aorta. These patients may be asymptomatic and they may have no associated lesions. In this form most commonly associated anomalies involve the great vessels, namely patent ductus arteriosus, aortico-pulmonary window or coarctation of the aorta. If there is a single vessel supplying the right lung, this vessel can be anastomosed with the main pulmonary artery by using a graft.[3] Embryologically it has been thought that the anomaly results from the delay of the right sixth branchial arch to fuse with the left before the aorta and pulmonary artery separate from the truncus arteriosus, so that the right sixth arch is left in connection with the ascending aorta and supplies the right lung.

Absence of the left pulmonary artery is rare.[4] The most striking feature of this type of anomaly is that it is almost always associated with abnormalities of the bulbus cordis, therefore defects of the upper part of the interventricular septum and right ventricular infundibulum are encountered. Embryologically it can be explained by Bremer's theory on the development of pulmonary arteries. According to this theory the asymmetry of the pulmonary arteries results from the absorption of the proximal part of the left sixth branchial arch and any defect in the absorption of this part causes defective lesions in the bulbus cordis.[4]

Right aortic arch is also very commonly associated with the absence of the left pulmonary artery, an anomaly which does not occur when the right pulmonary artery is absent. As is commonly known in 20 to 25 per cent of cases of tetralogy of Fallot right aortic arch exists, but in the absence of the left pulmonary artery, right aortic arch is found in 60 per cent of the cases.[5]

As a result of bulbus cordis anomalies, the absence of the left pulmonary artery is very frequently associated with the tetralogy of Fallot.[6, 7] Asymptomatic cases have also been reported, but this is very rare.[8] The first case of an absent left pulmonary artery in conjunction with the tetralogy of Fallot was reported by Thomas in

1941.[9] We could find only 20 such cases reported in the literature, and in these cases the left lung was supplied by collateral bronchial arteries. It was not stated how many of them had separate single vessels arising from the descending aorta.

To our knowledge Taussig's case is the only one in which a single vessel was shown arising from the descending aorta, but no other defects were present. When the lung is supplied by a single vessel rather than collateral bronchial arteries, it seems to be more proper to call the situation «hemitruncus» as Taussig did.[10]

Our case showed the same basic anomaly, but an infundibular pulmonary stenosis, a large ventricular septal defect with right to left shunt and a considerable degree of overriding of the aorta were also present. Since the general condition of the child was fairly good and the abnormal vessel arising from the aorta functioned like a Blalock shunt, no surgery was considered necessary.

*Summary*

We have presented a case of hemitruncus associated with the tetralogy of Fallot and a brief review of the literature. The possibility of embryological development of the anomaly was discussed. When the absent pulmonary artery is replaced by a single vessel rather than collateral bronchial arteries, it seems more suitable to use the term hemitruncus.

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