Trunical Inversion with Biventricular Pulmonary Trunk and Aorta from Right Ventricle (Variant of Taussig-Bing Complex)

By Aaron B. Shaffer, M.D., José F. Lopez, M.D., Irwin K. Kline, M.D., and Maurice Lev, M.D.

SUMMARY
A heart is described pathologically in which the aorta emerged from the right ventricle and was not related to the ventricular septal defect, while the pulmonary trunk emerged from both ventricles, but mostly the right, and was related to the ventricular septal defect. This Taussig-Bing arrangement of vessels was coupled with the presence of the aortic orifice to the left and the pulmonic orifice to the right, which is an inverted position. The anatomic concept of inversion is an abnormality in position from the standpoint of laterality. A careful study of the conal regions of the left and right ventricles showed that those regions were not inverted. Therefore, this represents a case of Taussig-Bing complex with exclusively trunical inversion, which is unique. This may be explained on the basis of opposite metamic contribution to the development of the truncus.

Additional Indexing Words: Congenital heart disease Transposition of the great vessels Truncus Double-outlet right ventricle

We have recently studied a heart with biventricular origin of the pulmonary trunk, in which the aorta arose from the left side of the right ventricle anterior to the pulmonary trunk, which arose from the right side of the right ventricle posteriorly. The heart was in levocardia (normal position) with all organs in situs solitus. The atria and inlets of the ventricles and coni were in normal position. Thus, there was isolated inversion of the aorta and pulmonary trunk, which is unique; hence, the present report.

Clinical Review
The subject was a 3-month-old white female infant, with the following diagnoses, based on clinical, hemodynamic, and angiographic findings: nearly complete transposition of the great vessels with ventricular septal defect, bi-directional but predominantly left-to-right shunt, pulmonary hypertension, “infantile” or “transitional” coarctation of the aorta, and patent foramen ovale. The aortic root was found to be anterior to the root of the pulmonary trunk; however, the usual laterality of the ascending aorta was not noted during life. It could have been recognized by an unusual course of the catheter in the ascending aorta in the frontal plane. An aortogram in this projection was not obtained. In view of the precarious condition of the patient, surgical correction of the coarctation was recommended. At surgery (November 1961), a large patent ductus arteriosus was found to join the left pulmonary artery to the aorta distal to the coarctation. The coarctated segment of the aorta was re-
Figure 1

*Diagrammatic sketch of the anterior view of the unopened heart made from the opened heart. SVC = superior vena cava; PT = pulmonary trunk; and A = aorta.*

moved with an end-to-end anastomosis, and the patent ductus arteriosus was also resected. The patient did well initially, but succumbed on the second postoperative day.

**Pathological Examination**

Aside from the findings in the heart, the pathological diagnosis was marked pulmonary atelectasis (bilateral). The viscera were all in normal position.

**Heart**

Estimation of chamber size and muscle mass of chambers was made by the method of Lev and associates. The heart was enlarged and weighed 59 g (normal, about 28 g). The base-apex axis pointed toward the left and down. The apex was formed by both ventricles. Two arterial trunks emerged from the base: a large pulmonary trunk was situated posteriorly and to the right, and a smaller aorta anteriorly and to the left (fig. 1). The relationships of the atria, inlets of the ventricles, and coni were normal.

The right atrium was normal in size and its wall was of normal thickness. This chamber received the superior and inferior vena cavae and coronary sinus in a normal manner; the eustachian and thebesian valves were normal. The limbus was well formed, although small in extent, and the foramen ovale was obliquely patent, measuring 0.1 to 0.2 cm in greatest dimension. The endocardium of this chamber

Figure 2

*(Left) Right ventricular view showing emergence of pulmonary trunk, (Right) Diagrammatic sketch of left panel. PT = pulmonary trunk; A = aorta; D = ventricular septal defect; TV = tricuspid valve; P₁ = first parietal band; and P₂ = second parietal band. Arrow points to the outflow tract into the aorta.*
was diffusely whitened. The tricuspid orifice was enlarged; the tricuspid valve was normally formed but presented increased hemodynamic change.\textsuperscript{2,3}

The right ventricle was enlarged and its wall was thickened. The inflow tract had the normal architecture of the right ventricle. The outflow tract however was abnormal (figs. 2 and 3). A septal band consisting of several components proceeded from the left lateral wall to the base of the aorta. Here it formed a wide muscular structure. This structure gave off a parietal band (first parietal band) which extended over the anterior wall of the right ventricle. Part of the latter anchored on the septum again in the lower part of the outflow tract. A second parietal band extended from the base of the pulmonary trunk over the region adjacent to the anterior leaflet of the tricuspid valve. Thus the outflow tract of the right ventricle was divided into two parts leading into the aorta and pulmonary trunk with a more distinct conus-like structure beneath the aorta. This outflow tract, although abnormal, was clearly that of a right ventricle. The endocardium of the outflow tract was thickened. The aortic orifice was somewhat larger than normal. The aortic valve was normally formed, with marked hemodynamic change and prominent hillock formation at the commissures. Its annulus was not related to the tricuspid or mitral annulus. The coronary ostia were situated in the posterior and right anterior sinuses of Valsalva. The right coronary ostium gave rise to the anterior descending coronary artery and to the right circumflex. The latter gave off its usual branches to the right ventricle and terminated in the posterior descending coronary artery. The posterior coronary ostium gave rise to the left circumflex coronary artery, which in turn gave off the ramus anterior ventriculi sinistri and the ramus obtusus ventriculi sinistri. The coronary veins were not dissected. The transverse aorta was smaller than normal and presented groups of sutures. The pulmonary trunk emerged mostly from the right ventricle but straddled the ventricular septum over a defect to be described later.
The pulmonary orifice was larger than normal, its valve was normally formed, and it presented markedly increased hemodynamic change. Its annulus was in part related to the tricuspid valve. It was separated from the mitral annulus by a muscular ridge. The two pulmonary arteries were given off normally. The ductus arteriosus had been patent but was tied off at surgery.

The left atrium was markedly enlarged and its wall was thickened. Its endocardium was diffusely whitened. The mitral orifice was enlarged, its valve was normally formed, and it presented marked hemodynamic change.

The left ventricle (fig. 4) was enlarged, but its wall was normal in thickness. The architecture of the inflow and outflow tracts resembled that of a normal left ventricle. Its endocardium was diffusely whitened. The ventricular septum at its base presented a defect measuring about 1 cm in greatest dimension. This defect was situated in the anterior part of the ventricular septum and was confluent with the left side of the mouth of the pulmonary trunk. From this defect, a triangular muscle band extended obliquely downward to the base of the posterior papillary muscle and sent a prong to the region of the anterior papillary muscle.

The anatomic diagnoses were as follows:

Taussig-Bing heart with truncal inversion
1. Hypertrophy and enlargement of the heart
   a. Right ventricular hypertrophy and enlargement
   b. Left atrial hypertrophy and enlargement
   c. Left ventricular hypertrophy and enlargement

2. Ventricular septal defect

3. Fetal coarctation with an adult component with patent ductus and surgical intervention (removal of adult coarctation and closure of ductus)

4. Abnormal architecture of the ventricular septum


Discussion

Hearts with biventricular origin of the pulmonary trunk with origin of the aorta from the right ventricle, with or without pulmonary stenosis, have recently been studied by one of us.\textsuperscript{4,5} A concept of what we call the Taussig-Bing spectrum of hearts (or double-outlet right ventricle of the Taussig-Bing type) was thereby developed. It was suggested that the Taussig-Bing heart be considered an anatomic entity, in which the aorta arises from the right ventricle and is not related to the defect, while the pulmonary trunk arises from the right or both ventricles and is related to the defect. In addition, there is a distinct musculature of the conal region of the right ventricle, in which a septal band group proceeds to the base of the pulmonary trunk and connects to the first parietal band which separates the aorta from the pulmonary trunk. A second parietal band proceeds downward from the aorta and is related to the anterolateral leaflet of the tricuspid. Accordingly Taussig-Bing hearts were classified as (1) right ventricular without overriding pulmonary trunk, (2) right ventricular with overriding pulmonary trunk, (3) intermediate, and (4) left ventricular.

In our case the pulmonary trunk overrides the defect but emerges mostly from the right ventricle while the aorta emerges exclusively from the right ventricle away from the defect.
TRUNCAL INVERSION

The aorta and pulmonary trunk arise from positions in the right ventricle opposite in laterality to those in the usual Taussig-Bing heart as described. This is accompanied by a change in the conus of the right ventricle, where the septal band is related to the aorta and the second parietal band is related to the pulmonary trunk. The first parietal band however continues to separate the aorta and pulmonary trunk.

Despite the alteration in muscular architecture of the conus of the right ventricle, we consider this to be related to other hearts with biventricular origin of the pulmonary trunk, with the aorta emerging from the right ventricle. We consider the alteration in muscular architecture as being related to the alteration in laterality of the aorta and the pulmonary trunk. This alteration in laterality of the vessels, together with the anterior descending coronary artery coming off the right aortic sinus of Valsalva, is the anatomic hallmark of inversion of the truncal, trunco-conal or bulboventricular regions.6-22

Inversion, anatomically, may be considered to be a disturbance in laterality pursuant to a certain axis. This axis is the longitudinal axis of the body when the arterial trunks are considered, and the longitudinal axis of the heart when the atria and ventricles are considered. It is well known that the atria, ventricles, or the trunco-conal areas may be selectively inverted. It is not known whether the coni or arterial trunks may each be inverted alone, separate from the other. In our case the atria and sinuses of the ventricles were not inverted. On the other hand, the bases of the two arterial trunks, derived from the truncus, were clearly inverted. A problem arose in the interpretation of the conus. The normal conus has a distinct right side with septal and parietal bands, and an abbreviated left side fused with the remainder of the left ventricle. Traditionally we consider a conus inverted when its distinct portion with septal and parietal bands lies in the left-sided chamber rather than in the right. This was not true in our case, in which the distinct portion, bearing facsimiles of the septal and parietal bands, lay in the right-sided chamber. Therefore we judge the conus not to be inverted.

A Taussig-Bing heart, as above defined, with trunco-conal inversion should have the pulmonary trunk straddling the interventricular septum, while the aorta emerges completely from the left-sided ventricle over a septal and parietal band; hence it would be similar to a corrected transposition. The aortic annulus, of course, would be anterior and to the left of the pulmonary annulus. This anatomic arrangement would not be physiologically the same as Taussig-Bing heart without inversion, or in our interpretation of the Taussig-Bing heart with truncal inversion, in both of which the aorta is in the line of systemic venous blood.

This anatomic concept of inversion may be expanded into a pathogenetic one, as Spitzer7 has done. We may be dealing with a reverse contribution of metameres as may occur in any one segment during the formation of the original single heart tube from its two-sided anlage.2 This inverse contribution of metameres in our case is assumed to occur only in the aortic bulb (truncal) area. The sino-atrial and ventriculobulbar contributions are presumed to be normal in this view, and the bulboventricular loop is normally formed. In this respect it may be recalled that the "aortic bulb" (truncus) is a distinct structure separate from the bulbus cordis.28 Together with the reverse metameric formation of the truncus is the unknown cause of transposition leading to biventricular origin of the pulmonary trunk, with the aorta emerging from the right ventricle.

Conclusion

A case is presented with biventricular origin of the pulmonary trunk, and origin of the aorta from the right ventricle, with truncal inversion. The aorta emerged from the right ventricle unrelated to a ventricular septal defect while the pulmonary trunk overrode the septum over a defect, but emerged mostly from the right ventricle. Altered septal and parietal bands were found in the right-sided ventricle. The aorta and pulmonary trunk
were inverted, while the atria and the sinuses of the ventricles were not inverted. Evidence is presented toward the concept that the conus was not inverted.

References


Truncal Inversion with Biventricular Pulmonary Trunk and Aorta from Right Ventricle (Variant of Taussig-Bing Complex)

AARON B. SHAFFER, JOSÉ F. LOPEZ, IRWIN K. KLINE and MAURICE LEV

_Circulation_. 1967;36:783-788
doi: 10.1161/01.CIR.36.5.783

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1967 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/36/5/783

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in _Circulation_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation_ is online at:
http://circ.ahajournals.org//subscriptions/