Anomalous Insertion of the Persisting Right or Left Ductus Arteriosus*

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Recently the authors encountered a rare anomaly of the ductus arteriosus in a 5 month old girl. Although the same anomaly had already been reported by some authors, embryological consideration of this case will also clarify the derivation of the anomalies which have the similar configuration to the present case, particularly of the absence of a unilateral pulmonary artery, and the purpose of this paper is to discuss the interrelation among these anomalies.

Many reports have been published hitherto about the various types of anomalies of the arch of aorta, but only a few papers had mentioned about the interrelationship among these anomalies, and most of these had discussed of their own case only. Since most anomalies of the arch of aorta are thought to be based upon the common embryological derivation, not to have the different origin in each case (Kasai, 1962), we must always pay our attention upon the various anomalies which seem to have the common features with one another in their vascular arrangement. Moreover, the present authors are of the opinion that for the purpose of understanding the essential nature of the various types of anomaly of the arch of aorta, informations on the behavior of the surrounding structures of the arch of aorta which are affected in position according to the transformation of aorta, especially of the ductus arteriosus, are of high importance and at the same time offer a key to the etiology of such anomalies.

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Findings of the present case (Figs. 1 and 2)

The present case was found at the dissection of a 5 month old female. The infant had been suffered from cyanosis and dyspnea before death, and was diagnosed the congenital heart disease. Absence of spleen was recognized by dissection, but the other remarkable malformations were not found except that of the heart and great vessels described below. The heart was normal in size and in

![Photograph showing the heart and great vessels of the present case. Ventral view.](image)

position, and the veins entering the heart were also normal. In the interior of the heart, the persistence of the primary common atrioventricular canal and the ventricular septal defect were observed, and the foramen ovale was widely patent. Therefore the two atria and the common ventricle had a free communication among them, forming the so-called ‘Cor triloculare biatriarum’. Neither the tricuspidal nor the mitral valve was present, and there was an atrioventricular valve, common to both sides of the heart, which consisted of anterior and posterior leaflets.

The pulmonary trunk was in normal position, but the outflow tract from the ventricle was completely obstructed, and then the pulmonary valves were not present. The blind end of the pulmonary trunk expanded forming the diverticulum-like enlargement, and was enclosed within the myocardial layer of the heart.

The ascending aorta arose from the common ventricle, coursing upwards to the right of the pulmonary trunk, and arched over the right bronchus forming the right-sided arch of aorta. Thereafter it bent to the left side of the midline, passing between the vertebral column and the esophagus, and continued to the descending aorta which was in normal position. Three branches were given off from the arch of aorta in the following order, that is, the left innominate artery, the right common carotid artery and the right subclavian artery. The left innominate artery was divided into the left common carotid and the left subclavian arteries. Therefore the branching from the arch of aorta was the mirror-image of normal, and this belongs to the so-called M-type aorta (Kasai, 1962).

The aspect of the ductus arteriosus in this case is of importance, and the present study was primarily designed to report on this point. An anomalous artery, measuring 5 mm. in diameter, was found connecting the left subclavian artery with the left pulmonary artery. The insertion

Fig. 2. Schematic representation of Fig. 1.
of this artery to the left pulmonary artery was near the bifurcation of the pulmonary trunk into the right and left pulmonary arteries. The other end attached to the left subclavian artery near its origin from the left innominate artery. From the view point of embryology and also from its topographic relation to the surrounding structures, it will reasonably be considered that this anomalous artery presents no more than the patent left ductus arteriosus. On the other hand, the right ductus arteriosus or the structures corresponding to it was not observed. The right recurrent laryngeal nerve hooked around the arch of aorta from right to left, but the origin of the left recurrent laryngeal nerve from the left vagus nerve was not easily decided, because the plexus-like communications among the recurrent laryngeal nerve and the cardiac branches were observed around the left ductus arteriosus. However the main stem of this nerve seemed to hook around the left subclavian artery backwards.

Discussion

As mentioned in the previous paper, it would be of interest to know how the ductus arteriosus is transformed accompanying the various anomalies of the arch of aorta. Although the numerous papers have been published concerning the anomalies of the arch of aorta, few papers had discussed on this point. Kasai (1962) had concluded that the ductus arteriosus usually remained unaffected in anomaly of the arch of aorta of any type, and that even when the arch of aorta was transpositioned to the right forming the so-called right arch of aorta the ductus arteriosus appeared in the left side connecting between the left pulmonary artery and the descending aorta passing over the left bronchus. This conclusion can be adapted in most types of anomaly of the arch of aorta, especially in those in adult. However the exceptional cases to this rule have rarely been reported by some authors, and the present case of this paper was also one of these rare cases. In these exceptional cases, the ductus arteriosus extended from the left pulmonary artery to the left innominate or subclavian artery coexisting with the right-sided arch of aorta which had the mirror image branching of normal arrangement, or in other cases the ductus arteriosus appeared in the right side connecting the right pulmonary artery with the right innominate or subclavian artery which arose from the normally left-sided arch of aorta.

When we analyze these various forms of the ductus arteriosus
from the viewpoint of embryology, the following four types can be classified (Fig. 3). In the first type, the left sixth aortic arch persists in its total length, and continues to the left dorsal aortic root, passing through the left dorsal aorta downwards. This is the most common feature of the ductus arteriosus, and that of the normal case is of course included in this group. Furthermore the ductus arteriosus in most of the anomalous cases of the arch of aorta is also contained in this category, because, as mentioned above, the ductus arteriosus usually remains unaffected against the transformation of the arch of aorta and connects between the left pulmonary artery and the descending aorta. In these cases other malformations are scarcely combined. See the previous paper by Kasai (1962) for the detail of the ductus arteriosus of this type. The second type is characterized by the fact that the left dorsal aortic root is interrupted on the way of development and as a result the arch of aorta is right-sided and the left sixth aortic arch (the ductus arteriosus) continues to the left innominate or subclavian artery as the present case of this paper. The vascular arrangement of the third type roughly shows the mirror image of the second type, that is, the ductus arteriosus is produced in the right side and extends from the right pulmonary artery to the right innominate or subclavian artery which arises from the normally left-sided arch of aorta. In the second and third types severe malformations are usually combined and the patients are tended to die at an early age. In the fourth type
the ductus arteriosus appears in the opposite side of the normally left-sided arch of aorta and connects between the right pulmonary artery and the descending aorta passing over the right bronchus. However the fourth type will not be treated here, because the case reports on this type are too small to discuss and the detailed descriptions are wanted. Besides the above mentioned forms of the ductus arteriosus, another one has to be added. It is the right ductus arteriosus connecting between the right pulmonary artery and the right-sided arch of aorta and therefore this type mostly appears in the case of dextrocardia or situs inversus. However this may be left out of consideration of the present paper. Accordingly the second and the third types of the ductus arteriosus will preferentially be discussed in the following.

In the second type of the ductus arteriosus an anomalous artery, which has been thought to be derived from the left sixth aortic arch corresponding to the left ductus arteriosus, connects between the left pulmonary artery and the left innominate or subclavian artery in company with the right arch of aorta. In the third type, the mirror image of the former, an artery which is regarded as the right ductus arteriosus arises from the right pulmonary artery and attaches to the right innominate or subclavian artery. Consequently both types are symmetrical in form. However, exactly speaking, only a different point between them will be the site into which the pulmonary end of the ductus arteriosus is inserted. From the embryological consideration by Congdon (1922) the ventral half of the left sixth aortic arch contributes to the formation of the pulmonary trunk rather than the left pulmonary artery, whereas the corresponding portion of the right sixth aortic arch becomes the beginning of the right pulmonary artery. Therefore the left ducts arteriosus in the second type will properly be inserted into the bifurcation of the pulmonary trunk, and the right ductus arteriosus of the third type will connect with the right pulmonary artery. In this paper, however, the insertion of the pulmonary end of the left ductus arteriosus was described as the left pulmonary artery not as the pulmonary trunk, to make a comparison with the right ductus arteriosus of the third type.

Generally it will be very difficult to consider the developmental processes of an anomalous artery, but there has been a general agreement on the point that the anomalous artery which connects between the pulmonary artery and the appropriate subclavian artery in each of the second and third types would be derived from the left and right sixth aortic arches respectively corresponding to the
ductus arteriosus of each side. Krause (1868) had already described such an anomalous ductus arteriosus in Henle's text-book of anatomy, and Edwards (1948) pointed out that the right sixth aortic arch could persist as a fibrous or slightly patent cord and deformed the arterial pattern. Recently Shane (1956) had published an excellent paper concerning the persistence of the right sixth aortic arch. Although Shane's material was pig and he discussed only of the right sixth aortic arch, he had illustrated the various forms of the anomaly which resulted from the persistence of the sixth aortic arch, referring to previously published reports on human fetuses or newborns. In that paper, he described the anomalous artery connecting between the right pulmonary artery and the right subclavian artery, the right pulmonary artery arising from the right subclavian artery and the right subclavian artery taking the origin from the right pulmonary artery. As he stated, these anomalies will properly be considered to be derived from the patency of the right sixth aortic arch. Although he did not mention on the persistence of the left sixth aortic arch, this paper would be the only one which have ever treated on anomalies of the sixth aortic arch systematically.

To identify the anomalous artery under discussion as the ductus arteriosus, the relation between this artery and the recurrent laryngeal nerve of the same side should not be neglected, but this point had scarcely discussed previously. Brenner (1883) had proposed an important rule in higher vertebrates that the recurrent laryngeal nerve hooked around the last aortic arch at the lateral side to the pulmonary artery. Consequently in order to identify the anomalous artery as the ductus arteriosus, the site around which the recurrent laryngeal nerve hooks upwards must be examined.

From the standpoint of consideration mentioned above, the authors could collect from the past literatures the following various configurations of the right and left ducti arteriosi.

(I) Persistence of the left sixth aortic arch connecting between the left pulmonary artery and the left subclavian artery—the second type of the ductus arteriosus of this paper (Fig. 4). In this type the arch of aorta was always right-sided, forming the mirror image branching of normal arrangement. Three types were subdivided in this group as follows.

a) The left ductus arteriosus connects between the left pulmonary artery and the left subclavian artery (Fig. 4a).

b) The left subclavian artery arises from the left pulmonary...
Fig. 4. Persistence of the left ductus arteriosus in company with right-sided arch of aorta. a) left ductus arteriosus connects between left pulmonary artery and left subclavian artery. b) left subclavian artery arises from left pulmonary artery. c) left pulmonary artery is absent and the compensative artery arises from left subclavian artery. Author's name is presented in each type.

Cailliot (1807)....2 cases cited from Krause (1868)
Rudolphi et al. (1818)
Gruber (1846)
Ozaki et al. (1959)....20 year-old female. Cor bilocular. Left closed ductus arteriosus connected between left innominate and left pulmonary arteries.
Hallman (1964)....4 day male infant with asplenia. Arch of aorta was interrupted between right common carotid and right subclavian arteries. Right ductus arteriosus was patent and continued to descending aorta.
Edwards et al. (1965)....13 month old girl with Fallot's tetralogy.
Kasai et al. (1967)....Present case of this payer.

Brenner (1883)....Right closed ductus arteriosus connected between right pulmonary artery and arch of aorta. Origin of left subclavian artery was also closed which corresponded to left ductus arteriosus, and communicating branch existed between intercostal artery and left subclavian artery.
Ghon (1908)....4.5 month old girl. Right closed ductus arteriosus connected between right pulmonary artery and arch of aorta.
Barger et al. (1956)....6 day male infant. Right patent ductus arteriosus connected between right pulmonary artery and arch of aorta.
Kleinerman et al. (1958)....Full-term boy. Arch of aorta was interrupted between right common carotid and right subclavian arteries. Right patent ductus arteriosus continued to descending aorta.
Edwards (1960)....Fallot's tetralogy.

In this case, left pulmonary artery is absent and compensative artery arises from left subclavian artery. See our previous paper (Kasai et al. 1967) for the detailed findings of this type and the authors who reported of this anomaly.
Fig. 5. Persistence of the right ductus arteriosus in company with left-sided arch of aorta. a) right ductus arteriosus connects between right pulmonary artery and right subclavian artery. b) right subclavian artery arises from right pulmonary artery. c) right pulmonary artery is absent and the compensative artery arises from right subclavian artery. Author's name of each type is also presented.

Breschet (1826) .... cited from Brenner (1883) and Krause (1868).
McCullough et al. (1944) .... cited from Blalock (1948)
Kelsey et al. (1953) .... 10 month old with dextrocardia. Left closed ductus arteriosus connected between left pulmonary artery and arch of aorta.

Heyfelder (1829) .... cited from Krause (1868)
Shapiro (1930) .... cited from Shaner (1956)
Evan's (1933) .... 3 day old male. Arch of aorta was interrupted between left common carotid and left subclavian arteries. Left patent ductus arteriosus continued to descending aorta.
Barger et al. (1954) .... Newborn female. Arch of aorta was interrupted between left common carotid and left subclavian arteries. Left patent ductus arteriosus continued to descending aorta.
Kleinerman et al. (1958) .... Full-term female. Arch of aorta was interrupted between left common carotid and left subclavian arteries. Left patent ductus arteriosus continued to descending aorta.

In this case, right pulmonary artery is absent and compensative artery arises from right subclavian artery. See our previous paper (Kasai et al. 1967) for the detailed findings of this type and the authors who reported of this anomaly.
artery near the bifurcation of the pulmonary trunk (Fig. 4b). The beginning of this artery are suspected to be derived from the left ductus arteriosus.

c) The left pulmonary artery is absent and the pulmonary trunk continues to the right pulmonary artery only. Instead of this, an anomalous artery appears arising from the left subclavian artery and enters the left lung (Fig. 4c). This compensative pulmonary artery is thought to be derived from the left ductus arteriosus. The present authors had already discussed on this anomaly in the previous paper published in 1967.

(II) Persistence of the right sixth aortic arch connecting between the right pulmonary artery and the right subclavian artery—the third type of the ductus arteriosus of this paper (Fig. 5). In this type the arch of aorta was normally left-sided. This type was also subdivided into the following three types.

a) The right ductus arteriosus connects between the right pulmonary artery and the right subclavian artery (Fig. 5a).

b) The right subclavian artery arises from the right pulmonary artery. The beginning of this artery would be derived from the right ductus arteriosus (Fig. 5b).

c) The right pulmonary artery is absent and the pulmonary trunk continues to the left pulmonary artery only. An anomalous artery arises from the right subclavian artery compensating the absent right pulmonary artery and enters the right lung (Fig. 5c). The beginning of this anomalous artery is thought to be derived from the right ducts arteriosus. See our previous paper on this anomaly.

Consequently when the right or left sixth aortic arch remains patent in its total length and connects with the respective right or left subclavian artery, any type of the above mentioned three kinds of anomaly in each group will finally be produced. In other words, these three types of anomaly are suspected to have the same embryological derivation with one another.

As mentioned in our previous paper, various sources have been reported as to the origin of the compensative artery which takes the place of the absent pulmonary artery. The ascending aorta, the arch of aorta, the thoracic aorta and the innominate or subclavian artery have been predominantly described. Actually the right and the left innominate or subclavian arteries have been reported as the origin of the compensative pulmonary artery in 5 and 6 cases respectively. On the other hand, the development of these compensative
arteries has not been clarified satisfactorily. According to our present investigation, it would be properly ascertained that the compensative artery which arose from the right or left subclavian artery was derived from the persisting ductus arteriosus of each side.

Summary

An anomalous configuration of the left ductus arteriosus was observed at autopsy of a 5 month old female who was associated with the absence of spleen. The arch of aorta was transpositioned to the right and the branching from it was the mirror image of normal arrangement (M-type aorta). The left ductus arteriosus was patent and connected between the left pulmonary artery and the left subclavian artery.

Collecting the case reports concerning the anomalous insertion of the right and left ducti arteriosi from the past literature, the authors classified these into the following four types, taking the embryological derivation into consideration. In the first type, the left sixth aortic arch persists in total length and connects the left pulmonary artery (more correctly, the bifurcation of the pulmonary trunk after Congdon, 1922) with the arch of aorta or with the descending aorta passing over the left bronchus. Therefore not only the ductus arteriosus in normal aorta but also that in most anomalous cases of the arch of aorta are included in this type. See our previous paper by Kasai (1962) for the detail of this type. In the second type the persisting left sixth aortic arch connects between the left pulmonary artery and the left subclavian artery in company with the interruption of the left dorsal aortic root. In this type, the arch of aorta is always right-sided, and the branching from the aorta is the mirror image of normal. The third type roughly shows a symmetrical form to the second type, and then the arch of aorta is normally positioned. The fourth type is roughly symmetrical to the first. In this paper the type II and III were preferentially discussed, because the type I was minutely described in the previous paper and the type IV was left out of consideration because of the shortage of the case reports on this type.

Practically each of the second and the third types finally assumes the following three different forms in the vascular arrangement. 1) In the second type, the left ductus arteriosus connects between the left pulmonary artery and the left subclavian artery in company with the transposition of the arch of aorta to the right, and in the
third type, the right ductus arteriosus connects between the right pulmonary artery and the right subclavian artery which arises from the normally positioned arch of aorta. 2) The left (the second type) or right (the third type) subclavian artery arises from the pulmonary artery of the same side. 3) The pulmonary artery is unilaterally absent and the compensative artery arises from the subclavian artery of the same side.

Conclusively the above mentioned three types of anomaly in each type II and III seem to have the same embryological derivation with one another, and in other words, these three anomalies of each type would result from the persisting right or left ductus arteriosus.

References


