Rib Notching: With Reference to Its Various Causes and Diagnostic Implication of Cardiovascular Diseases

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Notching of the inferior borders of the ribs on the roentgenogram has been thought to be pathognomonic sign of coarctation of the aorta. This sign, however, has been known to occur in various other conditions in recent years. Although the various causes of unilateral or bilateral rib notching have been described, the diagnostic implications of this roentgenologic finding have not thoroughly been discussed. It would be probably important to discuss the causes and mechanisms of rib notching and its diagnostic implications, especially, to determine the surgical indication, on the basis of our presented cases and some references in the literatures.

The authors observed four cases of rib notching due to some cardiovascular diseases, in which coarctation of the aorta were excluded by means of aortogram and blood pressure curve in the aorta.

Case Report

Case 1 is a 21 year old man with an unusual variant of pulmonary arteriovenous fistula, namely, the one with a direct communication of the right lower pulmonary artery with the left atrium. There is marked, bilateral notching of the third to eighth ribs (Fig. 1). Pulmonary arteriogram in this case reveals that the majority of dye flows into the left atrium through the huge right lower pulmonary arteriovenous fistula and the opacification of the other lobar arteries is scarcely demonstrated except for the remains of dye in the left lower pulmonary artery. This patient shows marked cyanosis and the arterial oxygen saturation decreases to 70.8 per cent. The large amount of right to left shunt is presumed. The pulmonary vascular resistance is not calculated, but it is supposed to be low. In surgery, the dilated and tortuous intercostal arteries, the diffuse adhesions in the pleural space and the development of numerous small vessels which extend from the chest wall to the lung are demonstrated. It seems possible that the dilated intercostal arteries may contribute to the bronchial collateral circulation through these vessels in

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(Received for Publication, January 25, 1968)
compensation for the marked decrease of the effective pulmonary blood flow.

Case 2 is tricuspid atresia associated with transposition of the great vessels and pulmonary stenosis in a 36 year old woman. Notching of the left third to eighth ribs is demonstrated (Fig. 2). The patient shows cyanosis (arterial oxygen saturation is 87 per cent), polycythemia and digital clubbing. In her roentgenogram, the pleural adhesion to some extent in left side, and the reticulated vascular shadow over the periphery of the lung in same side are demonstrated. The pulmonary area generally appears clear, but the left middle and lower areas are more obscure than the right. The adhesion in right pleural space is scarcely demonstrated during the anastomosis of the superior vena cava to the right pulmonary artery. The cause of rib notching in this case is supposed to be similar to case 1.

Case 3 is a 27 year old woman of Takayasu's disease associated with the occlusion of left common carotid and left subclavian arteries, the severe narrowing of innominate artery with aneurysmal dilatation in the origin of this artery, aortic insufficiency with hypertrophic aortic valve, atherosclerotic dilatation of ascending aorta and aortic arch with the calcification, the tortuous coronary arteries, and the obstruction of the right upper pulmonary artery. The blood pressures are 250/0 mmHg at the right arm, 110/10 mmHg at the left arm, 120/70 mmHg at the lower limbs, 300/10 mmHg in the left ventricle, 280/60 mmHg in the ascending aorta, and 170/100 mmHg in the femoral artery. The retrograde aortogram reveals various vascular changes as mentioned above, and the narrowing of the descending and abdominal aorta (Fig. 3). The excision of the carotid body and the left upper thoracic sympathectomy are performed and her symptoms become improved thereafter. The ascending aorta and aortic arch are so hard as a stone and the pulsations in branches of aortic arch are hardly palpable during surgery. The adhesion in the pleural space is not present. Notching of right fourth to ninth and left fifth to ninth ribs is demonstrated, which is slightly in left side. The cause of rib notching in this case may be the result from the development of collateral circulations from the intercostal arteries to the upper limbs through the internal or external mammary and subscapular arteries and the sclerotic tortuosity of intercostal arteries them-

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selves. It may be expected that the rib notching in this case is also related to the hypertension with widened arterial pulse pressure.

Case 4 is a 41 year old man of aortic arch syndrome due probably to atheromatous origin associated with the occlusion of left subclavian artery, aortic insufficiency, atherosclerotic changes of the descending aorta and aortic arch with the calcification, and the narrowing of the abdominal aorta (Fig. 4 and 5). His blood pressures are 220/0 mmHg at the right arm, 150/10 mmHg at the left arm and 110/70 mmHg at the lower limbs. The notching of the left third to tenth ribs is demonstrated without the erosion of right ribs. The retrograde aortogram shows that the left third to tenth intercostal arteries are dilated and tortuous with the intensive narrowing of the abdominal aorta. The unilateral rib notching probably can be explained on the basis of the development of collateral circulations from the intercostal arteries to the left upper limb with the sclerotic tortuosity of the intercostal arteries themselves.

REPORTED CASE IN THE LITERATURES

The rib notching was first described as a roentgenologic sign of coarctation of the aorta by Roessler in 1928 and in the following year by Railsback and Dock. The rib notching in coarctation occurs characteristically at the third to eighth ribs and this sign is bilateral in most cases. In rare cases in which the coarctation is located proximal to the origin of the left subclavian artery, unilateral notching may occur on the right. In the coarctation, however, absence of this sign is not unusual. Holmes reported rib notching in 192 of 300 patients with coarctation. Reifenstein also reported rib notching in 75 per cent of adult cases with coarctation. In younger patients less than 5 year old with coarctation, this finding is uncommon, because the collateral circulation is not well developed. However, Martelle reported notching in 13 per cent of infants with coarctation under one year of age.

Recently numerous reports have been made of rib notching accompanied by any congenital cardiac malformation which was not associated with coarctation of the aorta. Batchelder and Williams, in 1948, reported a case of tetralogy of Fallot in which rib notching occurred on the left, and Campbell, in 1958, described two cases of tetralogy of Fallot, one of pulmonary atresia, and one of Ebstein's malformation. Moreover, Drexler, in 1964, observed one case with transposition of the great
vessels and pulmonary stenosis associated with the rib notching. Sherrick\textsuperscript{9} observed one patient with agenesia of a main branch of the pulmonary artery and rib notching on the involved side. A similar case has been reported by Felton\textsuperscript{10}, Stork\textsuperscript{11} and Prutzman\textsuperscript{12} described some cases of pulmonary arteriovenous fistula in which unilateral rib notching was present. Recently, Wilson\textsuperscript{13} has reported another similar case in which multiple rib notching was noted on the involved side. Good\textsuperscript{14} and Brain\textsuperscript{15} described rib notching in some cases of systemic-to-pulmonary arterio-arterial or arterio-venous fistulas. Hench\textsuperscript{16} reported a case of unilateral rib notching due to an intercostal arteriovenous fistula.

Rib notching accompanying an acquired cardiovascular condition has been observed as well. Aubry\textsuperscript{17}, in 1937, and DuSailant\textsuperscript{18}, in 1945, reported bilateral or unilateral rib notching in arteriosclerosis, especially those associated with aortic insufficiency, syphilitic aortitis and mitral stenosis associated with endocarditis. Weir\textsuperscript{19}, in 1956, observed a case of pulseless disease in which bilateral rib notching was present. It has been known for a long time that unilateral rib notching after the Blalock-Taussig operation may also occur on the anastomosed side. Such a case was first described by Kent\textsuperscript{20} in 1953. McCord\textsuperscript{21} discussed the various causes of rib notching in acquired condition and reported a peculiar case in which idiopathic superior vena caval obstruction was associated with rib notching.

Moreover, the other various conditions without any cardiovascular disease have been reported. The rib notching due to intercostal neurofibromatosis was observed by Holt\textsuperscript{22} and Hunt\textsuperscript{23}. However, Bavendam\textsuperscript{24} reported rib notching in 6 of 275,000 draftees in which no demonstrable cause was found on clinical histories and physical findings.

**CAUSES OF RIB NOTCHING AND ITS DIAGNOSTIC IMPLICATION**

Although rib notching is not pathognomonic of coarctation as previously stated in our presented cases and reported cases in the literatures, coarctation of the aorta is the most common cause of notching. Numerous studies have demonstrated the mechanism of rib notching in coarctation. It seems to be impotent that the collateral pathways in coarctation as well as in the other cardiovascular conditions are elucidated because of the special reference to the surgical indication. The collateral blood flow in coarctation arises from the subclavian arteries and goes down distal to the coarctation through the anastomoses between the intercostal arteries and the following pathways. The blood flows through the following three main pathways: (1) the scapular pathway, consisting of the transverse scapular, transverse cervical and superficial cervical arteries from the thyrocervical trunk, and the subscapular artery from the axillary artery; (2) the external thoracic pathway, consisting of the short and long thoracic branches of the axillary artery; (3) the internal mammary pathway. Usually the third to eighth ribs are involved in coarctation, because the first and second intercostal arteries arise from the superior intercostal artery, a branch of the costocervical artery. They do not flow directly into the descending aorta. The intercostal arteries below the tenth join the epigastric artery, a branch of the external iliac artery. They do not participate in the collateral pathways, referred to in the foregoing. Notching in coarctation may have important diagnostic implications in regard to the location and degree of the coarcted segment and the existence of associated anomalies.

Bilateral intensive rib notching in pulmonary arteriovenous fistula, in so far as we know, may be the first that have been reported in the literature. The cause of rib notching in pulmonary arteriovenous fistula ascribed to the presence of the dilated intercostal arteries carrying a systemic blood flow to the pulmonary arteriovenous fistula. However, notching in our presented case may be a result from the markedly augmented intercostal-bronchial collateral circulation as a compensation for the decreased effective pulmonary flow due to a large amount of the right to left shunt. The increased vascularity of chest wall through the presence of adhesion in the pleural space should be present in such cases. This may be important and useful references on determining the

*Japanese Circulation Journal Vol. 32, July 1968*
surgical indication. The cause of rib notching in the tricuspid atresia of our presented case may be presumed to be same as the one in the former case. The cardiac malformations associated with rib notching reported in the literatures are tetralogy of Fallot, pulmonary atresia, Ebstein's malformation, and transposition of the great vessels. None has been written about rib notching in the tricuspid atresia. In such congenital malformations, relative old-aged Morbus caeruleus and decreased pulmonary blood flow are unisonous. In tetralogy of Fallot, the collateral circulations through small vessels of pulmonary ligament from the musculo-phrenic artery and pericardiaco-phrenic artery from the internal mammary artery is present beside the direct bronchial arterial collateral pathway. On the roentgenogram of such cases, the closed net works of small vessels on the basis of the lung are frequently demonstrated. If the pleural adhesion is present from any cause, the intercostal-bronchial collateral pathway through this adhesion may develop and then some localized rib notching may occur. However, rib notching in tricuspid atresia of our presented case is localized on the left side. The pleural adhesion on the left side is no more than a guess on her roentgenogram, and a slight adhesion in the right thorax and the considerable development of vessels through this adhesion are demonstrated during surgery. If the pleural adhesion and the development of vessels between the lung, pleura and chest wall are diffuse and massive, rib notching may be intensive. Moreover, it seems to be important for surgery that the divesting of the diffuse adhesion may not be easy. Therefore, the indication for the shunt operation in such cases should be careful to determine. Moreover, the pulmonary resistance may be high because of increased bronchial collateral flow, if the collateral blood supply is pretty through this diffuse adhesion.

On the contrary, cause of rib notching in any aquired cardiovascular conditions seems to be complicated. The mechanism of notching in the pulseless disease (Takayasu's disease) of our presented case may be, as Drexler and Boone reported, due to the augment of the collateral flow to the subclavian or axillary arteries through the intercostal arteries; in other words, the collateral pathway is the one same as in coarctation, but the blood flow is in the reverse direction. Rib notching following the Blalock-Taussig operation is the one same as seen in this condition. Although rib notching accompanying pulseless disease has been usually explained on the basis mentioned in the foregoing, the other cause may be related with our own case by which rib notching may be produced more extensive in right side (non-occluded side in subclavian artery; the blood pressure in the right arm is 250/0 mmHg) than in left side (subclavian is occluded). This hypertension with widened arterial pulse pressure may be connected with the cause of rib notching. Moreover, rib notching should occur in aortic arch syndrome from atherosclerosis. No such a case has been reported in the literature as shown in our presented case.

Other causes of rib notching also have been reported in cases of superior vena caval syndrome, intercostal neurofibromatosis, idiopathic rib notching and lesions of ribs themselves. However, this subject is not dealt with in this report.

Summary

Rib notching following various congenital or acquired cardiovascular diseases is discussed in connection with its causes and diagnostic implication on the basis of our own cases and reported cases in the literature. This sign was widely accepted as pathognomonic of coarctation of the aorta, but it occurs in various conditions as the authors point out in this report. Moreover, it is revealed that rib notching may be a result from the markedly augmented intercostal-bronchial collateral circulation as a compensation for the decreased effective pulmonary flow due to a large amount of the right to left shunt, if the adhesion in the pleural space would be present in some congenital malformations with decreased pulmonary blood flow and relative old-aged Morbus caeruleus. This finding is considered an important reference on determining the surgical indication in these conditions in regard to the cause, location and degree of rib notching.
REFERENCES


Japanese Circulation Journal Vol. 32, July 1968