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New Approach to Dysphagia Lusoria due to the Aberrant Right Subclavian Artery

by

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INTRODUCTION

The term "dysphagia lusoria" means dysphagia caused by *Lusus naturae*, that is, nature's play or ein Spiel der Natur in German. This term was coined by BAYFORD[4], an English anatomist, in 1789, and applied to a case of progressive dysphagia which lasted all the patient's life beginning in infancy, and in which BAYFORD discovered at autopsy an aberrant right subclavian artery. This is probably the most common abnormality of the aortic arch. This artery arises, not as usual from the innominate artery, but from the posterior aspect of the extreme left side of the aortic arch beyond the origin of the left subclavian artery and courses obliquely upward, usually passing between the esophagus and the spine, to reach the right apex of the chest. In later years other authors, especially AUTENREITH, have extended the term to apply to a syndrome, primarily dysphagia, resulting from anomalies of the aorta and its branches. More recently, because of the diversity of these anomalies and the inconstancy of dysphagia, the more exact term "arteria lusoria" has been proposed.

Patients with this anomaly rarely experience any symptoms. When there is a predominance of dysphagia or respiratory symptoms, the treatment should be a surgical one. Since GROSS carried out the first operation successfully on a four-month-old baby on December 26, 1945[9], a rather simple method of operation has been prevailing, in which the anomalous artery is ligated and divided close to its origin, because these patients vary in age from three weeks to six years. Recently, we encountered two cases of this anomaly in which dysphagia became troublesome in the latter part of life, and we were able to treat them successfully by using our newly designed method of operation whereby we added the blood vessel anastomosis to the original one. As these cases are the first ones successfully treated by surgery in Japan, we hope to report on our experiences with them, including some comments on literatures.

CASE REPORTS

Case 1. K. T., a 65-year-old woman.

Chief complaint: Dysphagia.

Family history: Her father died of apoplexy, and her mother died of uterine cancer.

Past history: Fifteen years and ten years before admission she had undergone hysterectomy for uterine myoma, and right nephrectomy for pyelonephrotic contracted kidney, respectively. She suffered from jaundice during the last 4 years and was treated for 2 years.

Present history: For the past 3 years, she complained of food sticking in her throat and was obliged to swallow with an effort. This trouble gradually became intensified 3 months ago, but she was still able to eat 2 bowls of rice at each meal. She was admitted to our clinic May 22, 1962.

Physical examination: Stature, moderate; well nourished; pulse 60, regular; character of arterial wall, sclerotic; blood pressure, right 120/80 mmHg, left 118/80 mmHg; respiration, quiet, regular; no stridor, face a little swollen, chest shape was not abnormal. There were no important variations from the normal in the heart, lungs and abdomen. No clubbing of fingers and toes.

Laboratory examination: Erythrocytes 377×10^4, leucocytes 6700, Hb 75%, hemogram; neutro. metamyelo. 0.5%, stab. 48.5%, seg. 2n. 11.5%, eosino. 2.0%, baso. 0.5%, mono. 7.5%, small lympho. 29.5%, TP 7.0g dl, MBSR 42mm, Wa. test of serum (−), feces Benzidin test (+). In urine sediment a small number of leucocytes and epithelia of bladder was demonstrated. There also were Enterococcus and Escherichia coli in urine, which showed sensitivity to all kinds of antibiotics other than penicillin. Liver function test; icterus index 4, Co. R R_3, Cd. R R_7, thymol turbidity test 1-2, BSP within 30 min. less than 5%; NPN in serum, 26.8 mg/dl, Na 144.9mEq 1, K 4.29mEq 1, Ca 4.16mEq 1, Cl 108.5 mEq/1: Clearance test of kidney; eff. RPF 268cc/min., eff. RBF 394 cc/min., GFR 68.5cc min., FF 25.6%. Serial test of dilution and concentration abilities of kidneys:

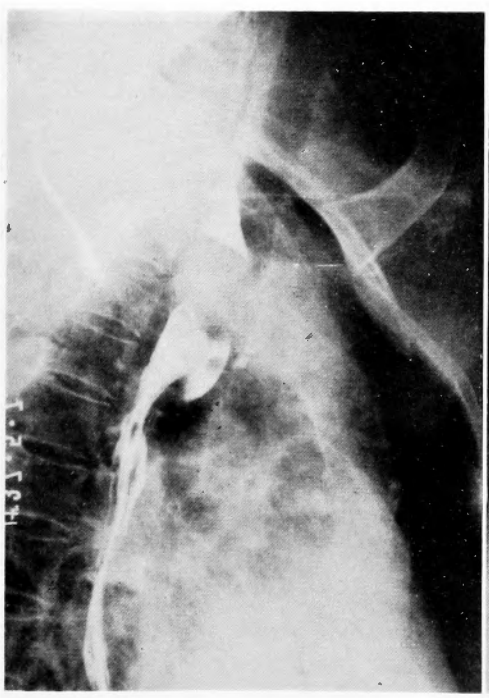

Fig. 1 Esophagogram of case 1.
Right anterior oblique projection.

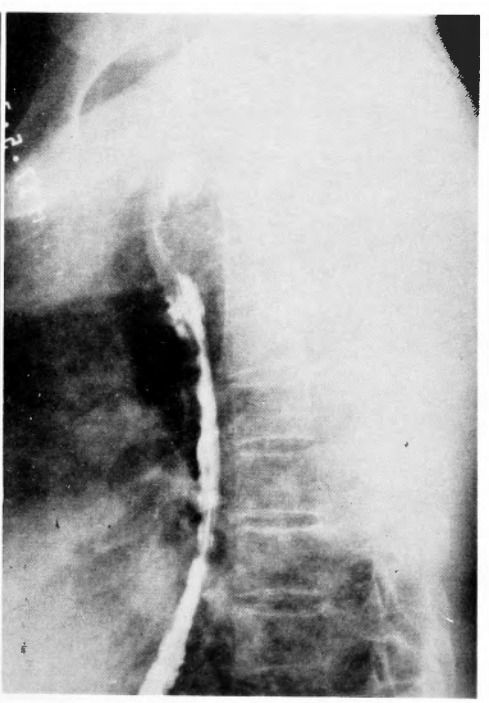

Fig. 2 Esophagogram of case 1.
Left anterior oblique projection.

The amount of urine was scarce for the initial 2 hours. ECG no abnormality, ventilation function of lungs; VC 2020cc,%VC 92%, MBC 60.5 1/min., %MBC 116%, %VEMS 83%.

Roentgenogram of the chest showed an enlargement of left first arch. The roentgen examination with barium swallow revealed a rounded indentation, over 3 cm in diameter and with sharp borders, in the posterior wall of the esophagus, at the level of the third and fourth thoracic vertebras in the right anterior oblique position and at the level of the second and third thoracic vertebras in the left anterior oblique position. The esophagus was displaced a little forward at that point, but the mucosal folds and the peristaltic activity were preserved normally (Figs. 1, 2).

From these findings, we performed an operation, under the erroneous diagnosis of leiomyoma, in the upper third of the thoracic esophagus.

The first operation (June 1,1962). Under intratracheal GOE intubationsanesthesia, anterolateral approach was made through the left side of the chest in the left recumbent position, entering the pleural cavity through the fifth intercostal space. The azygos vein was doubly ligated and divided. The thoracic segment of esophagus was totally dissected, but no abnormality of the esophagus was demonstrated. However, there was a pulsating strand of 0.8-1cm in diameter, running obliquely upward from the distal portion of the aortic arch reaching the right apex of the chest, whose originating portion from the aortic arch showed considerable dilatation. Consequently, this strand was confirmed to be the aberrant right subclavian artery. We were on the point of ligating and dividing the anomalous artery as the usual surgical procedure in such a case, but we gave it up for fear of the possible occurrence of circulation disturbance in the right arm, because the patient was very old. A catheter was introduced into the pleural cavity for continuous negative suction drainage. The chest was appropriately closed.

Postoperative course of the first operation. Although NPN value of the serum reached 33.3 mg/dl on the 3rd postoperative day, the postoperative course was uneventful and the drainage catheter was removed on the 3rd postoperative day. The wound healed per primum.

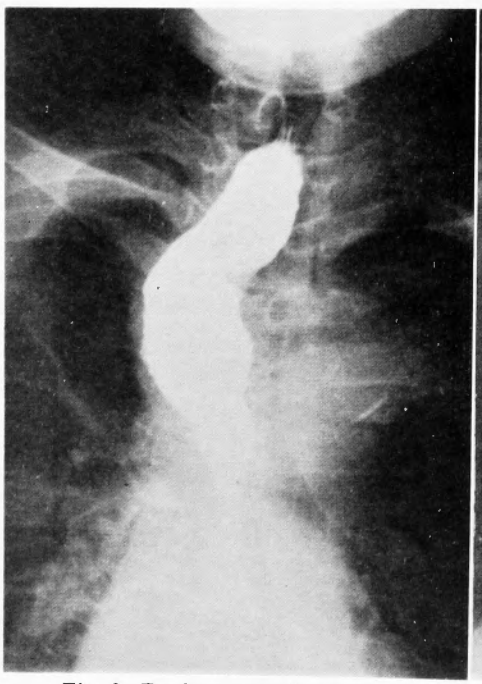

Fig. 3 Esophagogram of case 1. Anteroposterior projection.

Fig. 4 Angiocardiogram of case 1.

Then the roentgen examination of the esophagus with barium swallow was repeated. It revealed the same findings as the preoperative one in the oblique projection, but in the anteroposterior projection, the esophagus was displaced slightly to the right, and a typical oblique defect of the esophagus, running from the left caudal to the right cranial, was observed at the level between the second and the fourth thoracic vertebras. Angiocardiography revealed an abnormal course of the right subclavian artery and a considerable dilatation of its first portion near the aortic arch. However, no anomaly of the aorta and its branches were demonstrated (Figs. 3, 4).

As the patient still complained of dysphagia, we decided to perform a radical operation on this anomaly. No side-effect was observed after the right common carotid artery was occluded for about 20 minutes by MATAS's test.

The second operation (July 3, 1962). Under intratracheal GOF intubationsanesthesia, skin incision was performed in the right cervical and the anterior thoracic regions, as shown in Fig. 5.

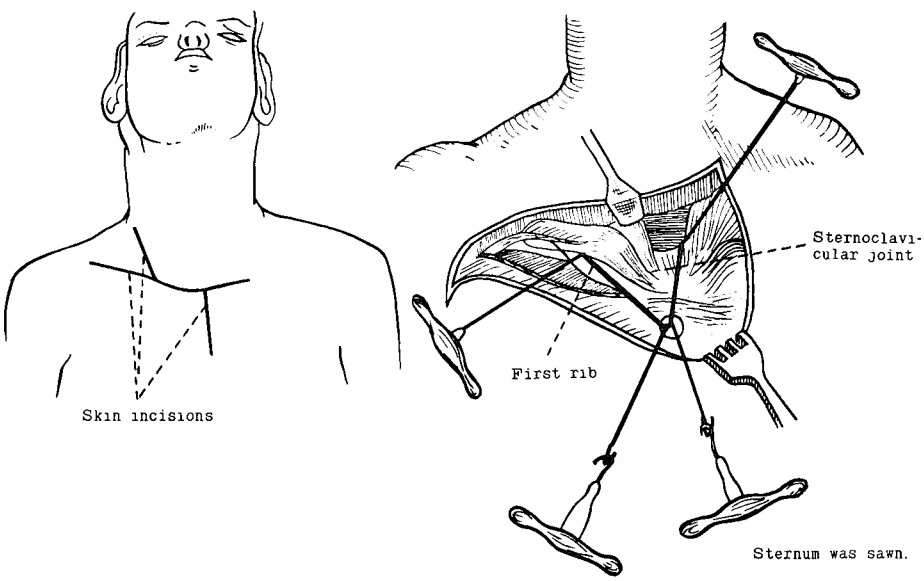

Fig. 5 Right sternoclavicular mediastinotomy by KILLIAN.

Dividing the right sternocleidomastoid and major pectoral muscles at their attached portion to clavicle, the right common carotid artery and the right internal jugular vein were dissected. Then the manubrium of sternum was divided according to the right sternoclavicular mediastinotomy by KILLIAN, as shown in Fig. 5. Its cranial segment was turned over to the right cranial side together with clavicle, and the cranial portion of the anterior mediastinum was exposed. Then, the trunk of the right subclavian artery was exposed by pursuing the region posterior to the right anterior scalene muscle and further dissecting toward the median cranial. At this time, the bifurcating portion of the innominate vein was carelessly damaged. Consequently, the right internal jugular vein was ligated and divided. And the right subclavian vein was almost completely ligated. Then, the trachea and the esophagus were dissected. The trunk of the anomalous artery was exposed in the retroesophageal region and dissected downward to reach the aortic arch. The anomalous artery showed a considerably firm adhesion to the surrounding tissues, probably owing to the first operation. Dividing the right internal mammary, vertebral and thyreocervical arteries close to their origins, the anomalous artery was doubly ligated and divided about 1 cm distal from the aortic arch and transposed to the right through the posterior aspect of the right internal jugular vein and the

right vagus nerve. Then an elliptical hole was made on the lateral wall of the right common carotid artery. The end-to-side anastomosis was carried out by hand without undue tension between the distal stump of the anomalous artery and the right common carotid artery. The caliber of anastomosed region was about 1 cm. The blood flow in the right common carotid artery was interrupted for about 25 minutes. The right radial pulse was fairly good after the release of interruption. The incised manubrium of sternum was sutured by means of 3 silver strings. The wound was closed, after a drain had been inserted into its superficial layer which was sucked under negative pressure. During this operation, 3600 cc of whole blood and 700 cc of 5% glucose solution were infused. The blood loss amounted to 3000 cc.

Histological examination of incised anomalous artery revealed the fibrinoid thickening and hyaline degeneration of intima and the intense fibrosis of adventitia.

Postoperative course of the second operation. She was placed into the oxygen tent for about 24 hours. Anticoagulant was not administered, but Kallikrein-depot was injected (1 amp. a day). The right radial pulse was good and regular, with the rhythm of 100–120 per minute. The maximal value of blood pressure ranged between 100–105mmHg in leg. No cerebral damage was induced. However, on the 2nd postoperative day the edema manifested itself in face, neck and arms, especially in the right shoulder and arm. She complained of pains in head and neck and vomited even a fluid. On the 4th postoperative day the edema had been intensified and the serous exudates flowed out from the wound. She complained of stridor and dyspnea. The right lung was short to percussion and moist râles were heard by auscultation. Because of this syndrome of superior vena caval obstruction, the inferior tracheotomy was performed and a large amount of intratracheal excretions was removed. On the 5th postoperative day, respiration became quiet and the edema decreased slightly. However, as she complained of difficulty in sleeping and swallowing, the LEVIN tube was left in the stomach for tube feeding. On the 8th postoperative day, a portion of the wound in the anterior

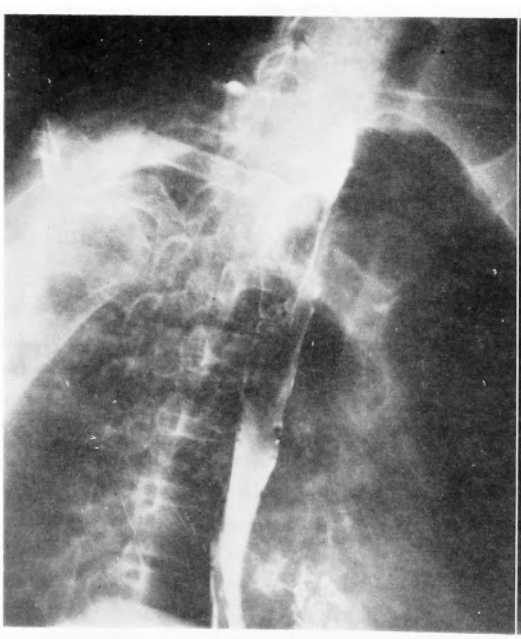

Fig. 6 Esophagogram of case 1, on the 260th postoperative day. Right anterior oblique projection. This shows complete disappearance of the indentation.

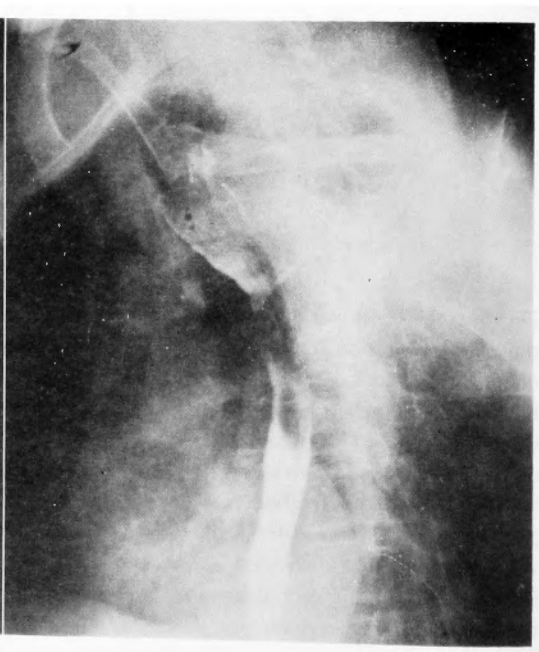

Fig. 7 Esophagogram of case 1, on the 260th postoperative day. Left anterior oblique projection. This shows complete disappearance of the indentation.

thoracic region was ruptured and a large quantity of bloody-serous exudates was discharged. And the syndrome of superior vena caval obstruction had diminished abruptly. The LEVIN tube was removed on the 15th postoperative day and the tracheal canula on the 20th postoperative day. However, as necrosis of the right sternoclavicular joint region occurred, possibly by the division of the right internal mammary artery which nourished this region, the wound in the anterior thoracic region was subjected to infection and incomplete healing. This wound was finally healed on March 16, 1963, after repeated incisions or scratches and sequestrotomy had been performed 3 times. She was discharged from the hospital in a very satisfactory state on March 16, 1963. At that time, she had had no trouble in swallowing. And regular pulsations could be felt in the right wrist. The blood pressure in the right arm was 119/80 mm Hg, identical to the value in the left arm. Barium examination of the esophagus showed it to be normal with no filling defect (Figs. 6, 7).

Case 2. S. N., a 67-year-old woman.

Chief complaint: Paroxysmal dysphagia.

Family history: Her mother died of cancer of the breast.

Past history: She had been suffering from ascariasis for about 10 years.

Present history: Since 5 years, she complained of food sticking in her upper substernal region after rapid swallowing of cold fluids, but could swallow within 5 minutes, if she hyperextended her head and had her back massaged. These bouts came on once or twice a month showing no aggravation. She consulted a doctor because of severe pains in the left neck and shoulder one month ago, and was diagnosed to be suffering from the tumor of the esophagus by roentgen examination. She was admitted to our clinic February 1, 1964.

Physical examination. Stature, moderate; well nourished, pulse 60, regular; character of arterial wall slightly sclerotic; blood pressure, right 162/88mmHg, left 158/85mmHg; respiration, quiet, 16, regular; no stridor; chest shape was not abnormal. There was no important variations from the normal in the heart, lungs and abdomen.

Laboratory examination: Erythrocytes 382×10^4, leucocytes 5800, Hb 75%, 12.1g/dl, blood platelets 16.2×10^4, Ht 38.5%, hemogram normal; TP 7.5g/dl, AI 52.5%, α_1 2.6%, α_2 10.5%, β 13.2%, γ 21.1%, A/G 1.11, bleeding time 1'0", clotting time 10'0", prothrombin time 17.5", MBSR 95.8 mm, serum cholesterol; total 202 mg/dl, ester 127 mg/dl, Wa. test of serum (−), urine; heat and acid test (+), feces, no abnormality; liver function test; icterus index 5, Co. R R_5, Cd. R R_5, thymol turbidity test 2-3, BSP within 30 min. less than 5%, PSP 15' 20%, 30' Σ30%, 60' Σ45%, 120' Σ55%, ECG; sinus bradycardia with nodal premature beat, left axis deviation; ventilation function of lungs; VC 2220 cc, %VC 100%, MBC 52.2 l/min., %MBC 96%, %VEMS 78%.

Roentgenogram of the chest showed the enlargement of left first arch. The roentgen examination with barium swallow revealed a rounded indentation of about 1.5 cm in diameter with sharp borders in the posterior wall of the esophagus, at the level of the third thoracic vertebra in the right anterior oblique position, and at the level of the second and third thoracic vertebras in the left anterior oblique position. The esophagus was displaced forward at that point, but the mucosal folds and the peristaltic activity were preserved normally. This indentation is best recognized in the left anterior oblique position. The anteroposterior projection showed an indentation of about 1 cm in breadth, running obliquely upward to the right cranial at the level of the third thoracic vertebra, just above the shadow of the aortic arch. Moreover, the esophagus was displaced backward with a possible filling defect and irregular mucosal folds in its anterior wall, of 3 cm in length, at the level of tracheal bifurcation (Figs. 8, 9, 10). The retrograde aortography demonstrated the aberrant right subclavian artery originating in the distal portion of the aortic arch, but no abnormality was observed in the course of the aorta and its major branches. However, both the abnormally thick left and the abnormally thin right vertebral arteries originated respectively from each side of the subclavian artery (Fig. 11).

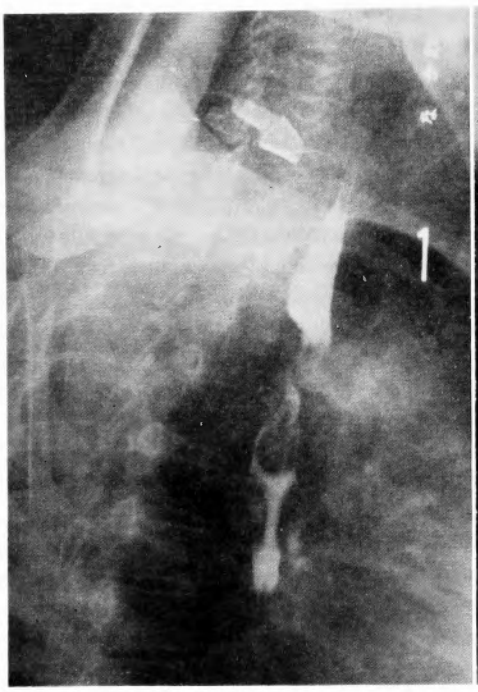

Fig. 8 Esophagogram of case 2.
Right anterior oblique projection.

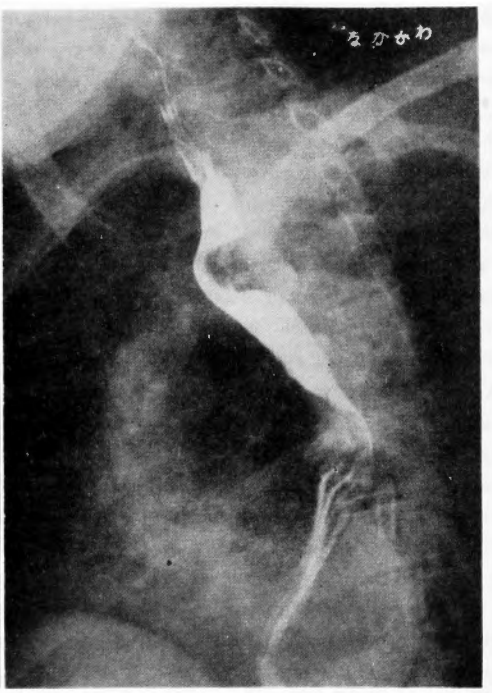

Fig. 9 Esophagogram of case 2.
Left anterior oblique projection.

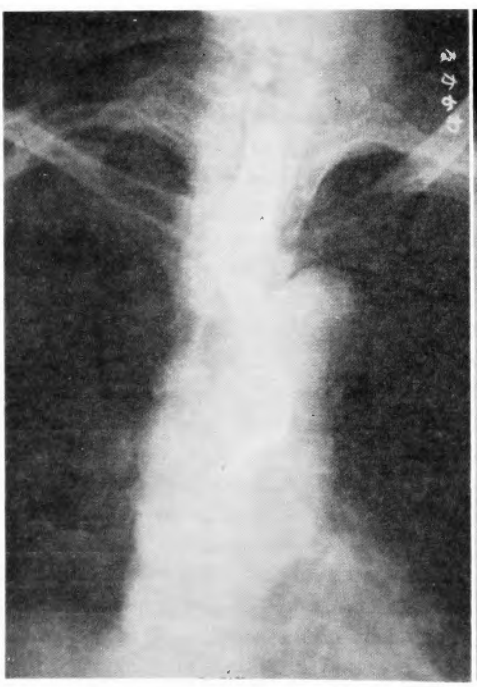

Fig. 10 Esophagogram of case 2.
Anteroposterior projection.

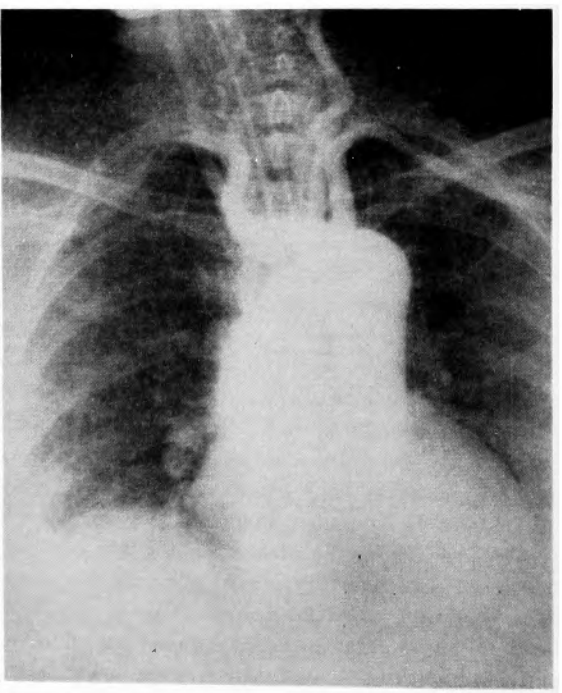

Fig. 11 Aortogram of case 2.

From these findings, this case was thought to be dysphagia lusoria due to the aberrant right subclavian artery. We attempted a radical operation, and to further investigate the mid-thoracic segment of the esophagus.

Operation (February 12, 1964). Under intratracheal GOFF intubationsanesthesia, anterolateral approach was made through the left side of the chest below the breast in the right semi-recumbent position, entering the pleural cavity through the 4th intercostal space. The 3rd and 4th costal cartilages were divided. This gave an excellent exposure of the upper mediastinum from its left lateral aspect. The aortic arch lay in a normal position. An anomalous artery of about 0.8 cm in diameter arose from the posterior aspect of the left side of the aortic arch, about 2 cm distal from the origin of left subclavian artery and then coursed upward to the right cranial side. This artery was extensively liberated from its bed after the longitudinal incision of the mediastinal pleura was made parallel to and behind the vagus nerve. This artery was confirmed to be the aberrant right subclavian artery, as it passed obliquely between the esophagus and the spine to reach the right apex of the chest and the interruption of its blood flow by compression with fingers resulted in the disappearance of the right radial pulse (Fig. 12). On the other hand, no abnormality was observed in the mid-thoracic segment of the esophagus. Therefore, the abnormal roentgenological findings prior to the operation in this region was thought to be due to compression by swollen mediastinal lymphnodes and adhesion. There was also a large pericardial cyst, the size of a man's fist, with the light-yellowish, transparent fluids as its contents, in the caudal portion of the mediastinum. This was extirpated. Then, 10 mg of heparin was injected into the distal portion of the anomalous artery, interrupting its blood flow by the traction of Nélaton's catheter looped around it. After 15 mg of heparin was administered intravenously, the anomalous artery was doubly ligated and divided near the aortic arch. The distal stump of the artery was permitted to retract to the patient's right and beyond the esophagus. The incision wound of the mediastinal pleura was closed mostly. A catheter was inserted into the pleural cavity for continuous negative suction drainage. The chest was appropriately closed.

The patient was then placed on her back. As the second step of the operation, another reversed L-shaped skin incision was made in the right cervical region, along the clavicle and the posterior border of sternocleidomastoid muscle. Platysma and omohyoid muscles were divided. Sternocleidomastoid muscle was displaced to the median side. And the right internal jugular vein and the right common carotid artery were dissected. Then the trunk of the right subclavian artery was exposed by pursuing the region posterior to the anterior scalene muscle. Dividing the right internal mammary, thyreocervical, transverse cervical and vertebral arteries close to their origins, the anomalous artery was obliquely divided a little distal from the origin of the thyreocervical artery. Then the right common carotid artery was transposed to the right through the posterior aspect of internal jugular vein and made an elliptical hole on its lateral wall. The end-to-side anastomosis was performed by hand between the distal stump of the anomalous artery and the right common carotid artery. The caliber of anastomosed region was about 1 cm. The blood flow in the right common carotid artery was interrupted for about 25 minutes, and that in the right subclavian artery for about one hour and 45 minutes (Fig. 13). The incised artery was 7.5cm in length, 6mm in caliber and 7mm in outside diameter. The right radial pulse was a little more feeble, as compared with the preoperative one, but showed a considerable magnitude. After a drain was inserted into its superficial layer, the wound in the cervical region was closed. During this operation, 600 cc of whole blood and 700 cc of 5% glucose solution were infused.

Histological examination of incised artery revealed a fibrinoid thickening and hyaline degeneration of intima.

Postoperative course. She was placed into the oxygen tent for about 22 hours. And 20 mg of heparin (Hepacarin) was injected intramuscularly thrice in every 3 hours. Thereafter, Sintrom was administered for 5 days (3-1/2 tablets a day). The radial pulse of both sides of arms were

Fig. 12 Operative findings of case 2. The view of the upper mediastinum from its left lateral aspect. Nélaton's catheter was looped around the aberrant right subclavian artery.

Fig. 13 Operative findings of case 2. The end-to-side anastomosis was done in the right cervical region between the distal stump of the anomalous artery and the right common carotid artery.

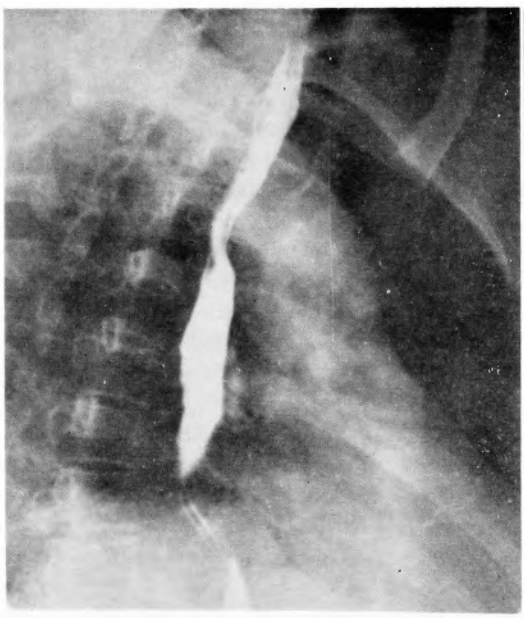

Fig. 14 Esophagogram of case 2, on the 19th postoperative day. Right anterior oblique projection. This shows complete disappearance of the indentation.

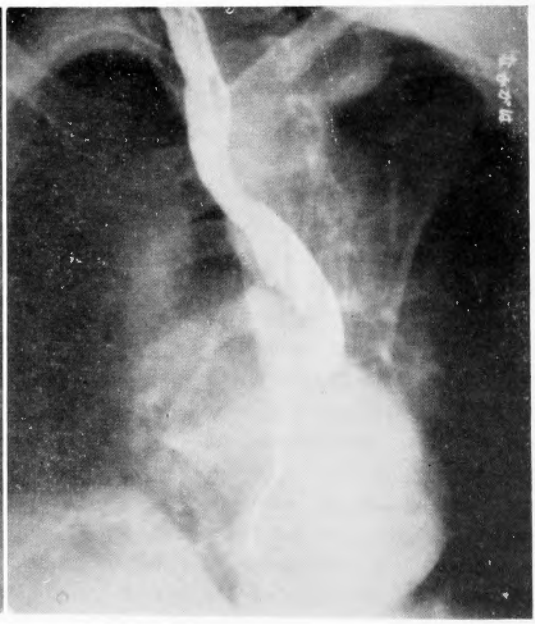

Fig. 15 Esophagogram of case 2, on the 19th postoperative day. Left anterior oblique projection. This shows complete disappearance of the indentation.

in the same degree 6 hours after the operation. Fortunately, no cerebral damage (anisocoria, hemiplegia etc.) was induced and the blood pressure ranged between the values of 130-140/80-90 mm Hg. A considerable amount of bloody pleural exudates was removed from the suction catheter. This catheter was removed on the 3rd postoperative day. As thoracenthesis showed still the accumulation of 220 cc of purely bloody exudates and the prothrombin time amounted to 93.5″, on the 6th postoperative day, 30 mg of Kaywan (vitamin K) was injected. Following this injection, the prothrombin time shortened (on the 7th postoperative day 31.0″, on the 8th postoperative day 18.0″) and the pleural exudates decreased in amount and became slightly yellowish and clear. No more exudate was obtained on the 22nd postoperative day. The postoperative course was uneventful and the wound healed per primum. Fluids and food were rapidly increased in amount. She took a solid diet on the 21st postoperative day, and she had no trouble in swallowing. Barium examination of the esophagus, on the 19th postoperative day, showed it to be normal (Figs. 14, 15). She was discharged from the hospital in a very satisfactory state on March 12, 1964 (Fig. 16). At that time, regular pulsations could be felt at the right wrist in the same degree as at the left, and the blood pressure was 142/88 mmHg at the right arm, while 138/88 mmHg at the left.

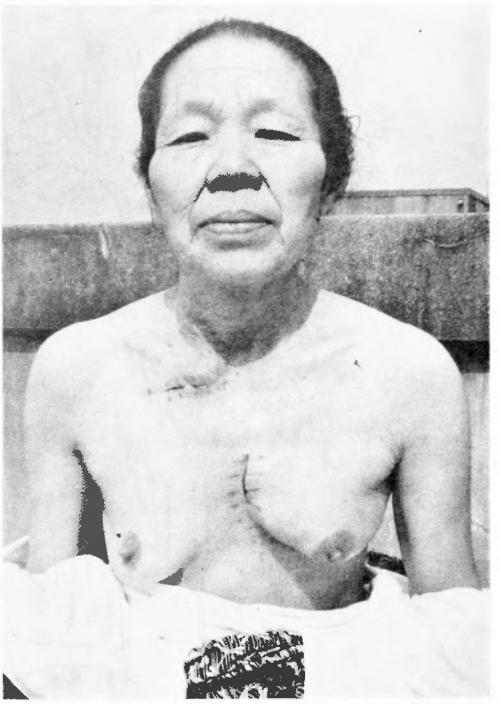

Fig. 16 Photo of case 2, when she left our hospital. Skin incisions which had healed per primum are observed.

COMMENT AND DISCUSSION

Classification. Although several variations of anomalies which may cause esophageal or tracheal compression have been described, dysphagia is usually due to one of three types: 1) double aortic arch, 2) right aortic arch with left ligamentum arteriosum or ductus arteriosus Botalli, and 3) aberrant right subclavian artery (Fig. 17).

Embryology and pathology. The arterial bulb which becomes the heart is continued in the early embryo by the two primitive ascending aortas which ascend toward the cephalic end of the organism along the ventral surface of the first branchial arch. These vessels curve toward the dorsal surface to form the first arterial or aortic arches, whence they turn toward the caudal to form the primitive descending aortas. During the second week of embryonal life the five anastomoses formed on each side between the ascending and descending portions of these aortas become the five primitive arterial or aortic arches. The aorta caudal to the branchial arches becomes fused as an unpaired dorsal aorta.

In the human being, the first, the second and the fifth arches become obliterated. The third arches on both sides persist as the proximal parts of the internal carotid artery. The left fourth arch persists as the definitive aortic arch, while most of the right fourth arch becomes the first portion of the right subclavian artery. The sections of primitive ascending aortas located between the origins of the third and the fourth arches become

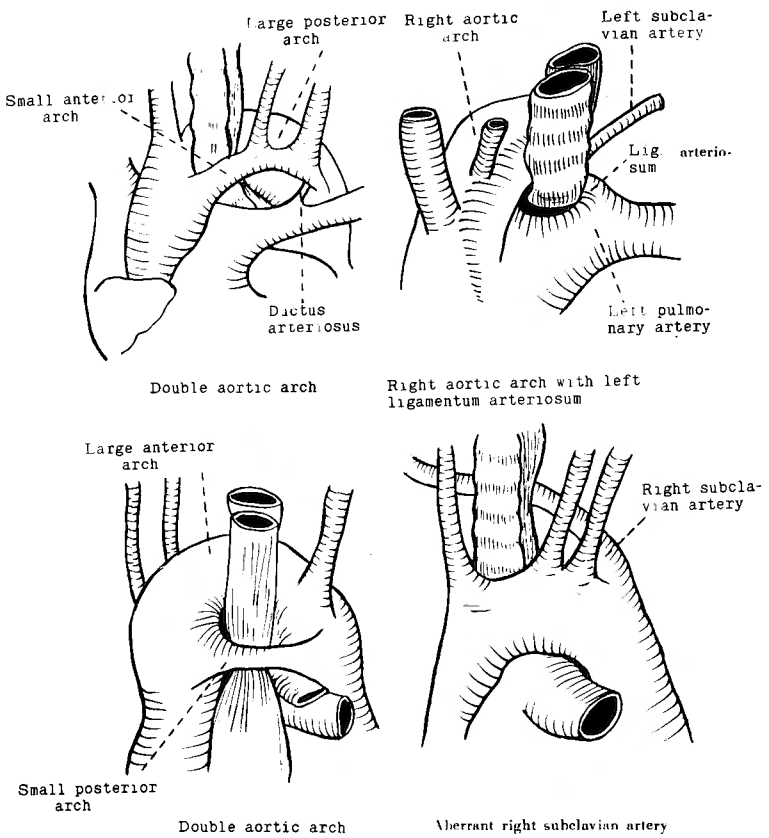

Fig. 17 Common vascular anomalies which may cause dysphagia lusoria.

the common carotid arteries. The distal portion of the right primitive ascending aorta becomes the innominate artery, while that of the left takes part in the formation of the aortic arch. Therefore, normally, the left ascending aorta, the left fourth aortic arch, and the left descending aorta form the final aortic arch of the adult. The left subclavian artery is a direct branch of the left descending aorta, while the right subclavian artery is formed by the right fourth aortic arch.

If the proximal rather than the distal part of the right fourth arch becomes obliterated in the transformation of the primitive aortic arches during the second month of embryonic life, the right dorsal aorta becomes obliterated not at the junction with the left dorsal aorta, but at its more proximal portion beyond the origin of the right subclavian artery. In such cases, the right subclavian artery retains its communication with the descending aorta, and arises not as usual from the right fourth arch, but from the diverticulum-like rest of the right primitive dorsal aorta, as the last branch from the aortic arch and crosses the spine anywhere from the sixth cervical to the fourth dorsal vertebra to reach the right arm. As the first portion of the aberrant right subclavian artery corresponds to the diverticulum-like remnant of the right primitive dorsal aorta, its wall is strikingly thin and sometimes shows an aneurysmal appearance. If the right aortic arch persists and the left arch becomes

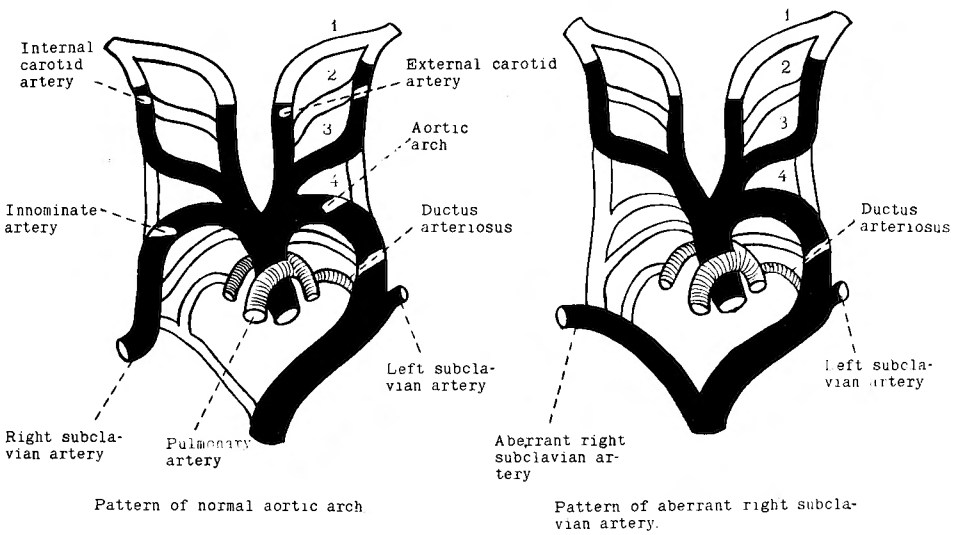

Fig. 18 Development of aortic arch and aberrant right subclavian artery.

obliterated, the exact mirror image of the above described arrangement may occur (Fig. 18).

The incidence of this anomaly ranged from 0.2% to 1.4% and it occurred frequently especially among the negroes[1]. According to HOLZAPFEL et al., 231 cases of this anomaly were reported in Europe before 1907 and according to ADACHI[1] seven cases were observed in Japan before 1924. There is a relatively high incidence of coexistent congenital heart diseases, such as patent ductus arteriosus or the tetralogy of FALLOT, and the coarctation of the aorta[5][16], and from 0.4% to 2% of all cases of congenital heart disease have this anomaly (Table 1). In 80-100% of the persons with this anomaly the anomalous artery courses behind the esophagus, but in 0-15% it passes between the esophagus and the trachea, and in 0-6% of the cases it passes in front of the trachea. In all of 36 cases reported by BAHNSON et al[3]., the artery was described as passing behind the esophagus. Among 133 cases in Europe reported by HOLZAPFEL, the artery passed behind the esophagus in 107 cases, between the esophagus and the trachea in 20 cases, and in front of the trachea in 6 cases. Summarizing the reports on autopsy of the cases in Japan, ADACHI[1]

Table 1. Incidence of the aberrant right subclavian artery

Author	No. of cases	No. of cases with the aberrant right subclavian artery	Materials	Incidence
Adachi, B. (1906-1918)	516	1	Japanese cadavers	0.2±0.20%
Quain	1000	4	Babies	0.4%
Holzapfel, G. (1899)	200			0.6%
McDonald, J. J. & B. J. Anson (1940)	1453	14		0.96%
Tomson, A. et al. (1889-1895)	500	5	English cadavers	1.0±0.44%
Szalowsky, J. (1888)	70	1	Fetuses	1.4%

has shown that the high incidence of such coexistent anomalies with this anomaly is accompanied by the failure of the right recurrent nerve, the right vertebral artery originating in the right common carotid artery or entering into Foramen transversarium at the unusually high level of cervical vertebra, the deviation or the overlap of thoracic duct, abnormal development of accessory hemiazygos vein and the persistence of left superior caval vein. The right recurrent nerve may enter into the larynx directly, loop around the vertebral or the inferior thyroid artery. According to ALLAN et al[2]., there is a simultaneous alteration in the course of visceral branches of the vagus, especially in the recurrent nerve, with no observable effect on its cardiac branches in this anomaly. However, sympathetic cardiac nerves are influenced by the alteration to a considerable degree. It was observed that the middle and inferior cervical cardiac nerves followed the aberrant vessel through its course to reach their distribution in the cardiac plexus.

Clinical manifestation. This anomaly is often asymptomatic. However, in a small percentage of these patients, paroxysmal or constant dysphagia may be troublesome or become aggravated by the extrinsic pressure on the esophagus; 1) when the artery is taut and stretches across the esophagus; 2) when the vessel becomes sclerotic in the latter part of life; or 3) when there is aneurysmal dilatation of the artery. Therefore, dysphagia may be present in babies and infants or in the aged. Some of these babies have no difficulty in swallowing milk or other fluids, but they encounter difficulties when solid or semisolid food is added to the diet. These vary from mild dysphagia to marked nutritional impairment. Usually these children have no respiratory distress, but there may be dyspnea (stridor, cough and suffocation), cyanosis, retraction of the head and sometimes pains in the upper thoracic segments and the arms. These symptoms are characteristically exaggerated during feedings, or in cases where the artery passes in front of the trachea. Irregularity of radial pulse and symptoms like those of the scalenus anticus syndrome might be produced. If the anomalous artery originates below the coarctation, the patient has hypotension of the right arm and legs, but hypertension in the left arm[16]. If the anomaly manifests itself in the latter part of life, there are episodic dysphagia, appearing without any obvious cause and usually associated with anxiety and a number of neurovegetative symptoms (vomiting, salivation, belching and lacrimation).

Diagnosis. If the infants complain of such distresses as dysphagia, stridor, cough, cyanosis etc. during feedings, or if the aged complain of gradually aggravated episodic dysphagia without obvious cause, it is necessary to examine them in detail, suspecting this disease. 1) In the roentgen examination with barium swallow, the lateral or oblique projection reveals a rounded indentation of rather small caliber with sharp borders in the posterior wall of the esophagus at the level of the third or fourth thoracic vertebra, just above the normal aortic compression. The esophagus is often displaced forward at that point, but the mucosal folds and the peristaltic activity may be preserved normally. This indentation is best recognized in the left anterior oblique position. If the anomalous artery passes between the esophagus and the trachea, the defect is in the anterior wall of the esophagus. In anteroposterior projection, this indentation runs obliquely upward to the patient's right, the direction and position of the defect being in a line from the distal part of the aortic arch to the right apex of the chest. This typical oblique de[illegible] is readily

seen if the barium is not too dense and if the amount swallowed is not too large. There is usually little or no dilatation of the esophagus above this area, but a delay in the passage of the barium is commonly observed (Figs. 1, 2, 3, 8, 9, 10), 2) The exact anatomical abnormality can be diagnosed by angiocardiography or aortography. It can demonstrate clearly the course of this anomalous artery. Frequently, there is an aneurysmal dilatation in the first portion of this artery, the shadow of which is sometimes overlapped by that of the aortic arch in the anteroposterior projection because this portion locates on the posterior surface of the distal portion of the aortic arch. The aortic arch is normal in size, position and direction (Figs. 4, 11). 3) The anomalous artery is demonstrated by the retrograde introduction through the right brachial artery of a radiopaque catheter. This catheter is seen lying along the oblique furrow in the esophagus caused by this artery. This method is less dangerous than angiocardiography. 4) The tracheo- and bronchogram show no abnormality.

Differential diagnosis. 1) Double aortic arch : It manifests itself clinically at an early age and primarily with severe respiratory symptoms, such as a crowing type of respiration with a marked inspiratory or exspiratory stridor, coughing, cyanosis and suffocation. Respiratory distress is often made worse during the process of swallowing, but dysphagia may not be noticed until they begin to eat a more solid diet, and it is not severe even when present. These babies have a high tendency to lie with the head in hyperextension. In the roentgenography with barium swallow, the oblique projection reveals a large posterior indentation of the esophagus located more in the caudal section as compared with the aberrant right subclavian artery. The anteroposterior projection reveals a moderate-sized indentation on both sides of the esophagus, nearly horizontal at the level of the third or fourth thoracic vertebra. In the visualization of trachea by Lipiodol, there is an indentation on both sides of the trachea in the anteroposterior projection and another one in the anterior wall of the trachea in the lateral projection, at the same level as in the esophagus. 2) Right aortic arch with left ligamentum arteriosum or left ductus arteriosus Botalli : Symptoms are similar to double aortic arch, but may be less severe and delayed in onset. In only 10% of the cases, there may be symptoms referable to swallowing. In the roentgen examination with barium swallow, the anteroposterior projection reveals a large indentation on right caused by the aortic arch and a narrow but deep indentation on the patient's left, which is made by ligamentum arteriosum. Left subclavian artery may arise on right and indent the posterior wall of the esophagus. The posterior type of right aortic arch usually displaces the esophagus to the left and anteriorly. In the tracheogram, there is a slight, a little longer defect on the right wall by aortic arch, and also a defect on the anterior wall by pulmonary artery and another one on the left wall by ligamentum arteriosum. In the case of aberrant right subclavian artery, one can see a normal aortic arch slightly to the left of the vertebral column, usually not at the site of the esophageal defect. And further, the defect is usually just above the aortic arch and is smaller than the posterior defect produced by a posterior right aortic arch. In all these cases, anterior displacement of the esophagus is less than that usually seen in posterior right aortic arch. 3) Benign intramural tumors of the esophagus, especially leiomyoma : They produce in profile a smooth, semilunar defect with intact mucosa and sharp borders, but the elasticity

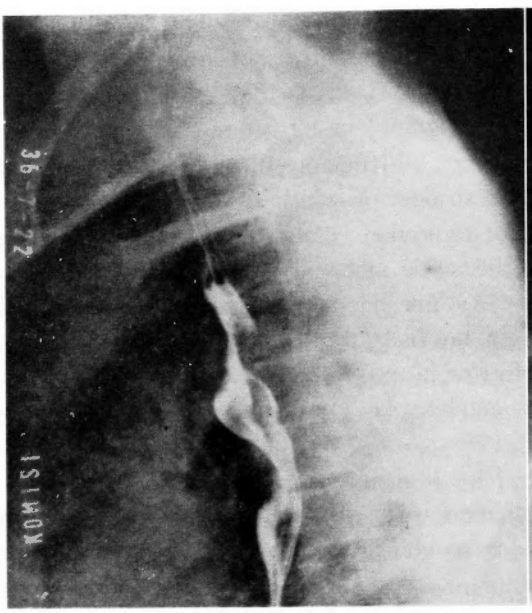

Fig. 19 Esophagogram of leiomyoma in the upper third of thoracic esophagus. Left anterior oblique projection, in profile.

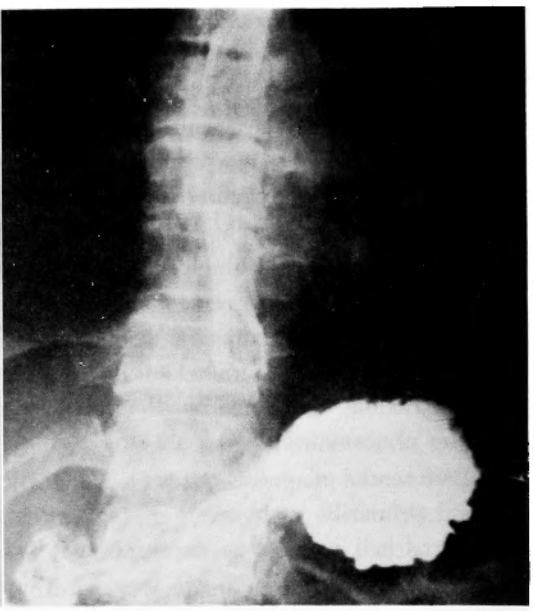

Fig. 20 Esophagogram of esophageal leiomyoma in the esophagogastric junction, in the direct plane.

and peristaltic movement of the esophageal wall may be preserved at that point. These findings quite resemble those in the aberrant right subclavian artery. However, in the direct plane, benign intramural esophageal tumor shows a rounded or elliptical central filling defect with sharp borders, as the mucosal folds are obliterated and causes the barium column to split or fork, while the aberrant right subclavian artery shows a typical defect running obliquely upward to the patient's right (Figs. 19, 20). Furthermore, the swollen mediastinal lymphnodes may give a similar roentgen picture to this anomaly. Anomalies of innominate or left common carotid arteries also cause compression symptoms of the trachea, but no dysphagia and roentgen examination of the esophagus in these cases show it to be normal.

Treatment. If the symptoms are mild, control of the type of food ingested and correction of bad eating habits may be all that is required. If these measures fail, or if the symptoms begin or become aggravated in the latter part of life owing to the arteriosclerotic or hypertensional changes, the difficulties can be overcome by surgical treatment. The surgical procedure to relieve dysphagia due to the aberrant right subclavian artery has been devised by GROSS and was successfully carried out on a four-month-old baby for the first time on November 26, 1945[9]. In the review by POSTLETHWAIT and SEALY[20], 16 cases of this anomaly were operated upon prior to 1961, and 14 cases survived this surgery. MAHONEY and MANNING[16] have described their experiences in performing an operation on two unusual cases of this anomaly. Usually the left side of the chest is chosen for the exposure of the anomalous vessel. According to GROSS, the operation attack has been through a left anterolateral incision with a transpleural approach and the chest was entered

throughout the length of the third intercostal space, and third and second costal cartilages were severed. He reported that this approach gave an extremely clear view of the anomalous artery. However, a midline sternotomy incision also provides easy access to all of the anomalies of the aortic arch or its major branches, thus eliminating the necessity of attempting to decide which side of the chest should be entered to provide the ideal exposure. If the patient's age ranges from three weeks to six years, as seen in the cases reported by GROSS, the surgical procedure for the alleviation of this syndrome is a rather simple one. It consists merely of exploration of the posterior mediastinum, freeing of the anomalous artery from its bed, doubly ligating and dividing it near its origin from the aortic arch. In fact, because the vessel is divided close to its origin, collateral circulation is so efficient that after a few days the irregular pulse usually reappears at the wrist and there is no need to worry about the blood supply of the arm. In general, the ligation of subclavian artery results in necrosis of the arm at the rate of about five per cent. But if the patients are over 40 years old, as seen in our cases, the incidence of necrosis may be higher than the former cases. As vascular surgery has made great progress recently, it is considered easy to perform not only the ligation and the division of the offending artery, but also the reconstruction of blood circulation in the right arm by end-to-side anastomosis between the distal stump of the anomalous artery and the right common carotid artery (Fig. 21).

From this point of view, in our first case, we tried to perform these two main operations by this procedure in the same field at once, using the right sternoclavicular mediastinotomy by KILLIAN. However, we encountered some difficulties. That is, as the first portion of the anomalous artery is located too far on the dorsal surface of the aortic arch in this field, its ligation and division were not so easy. And as the dissection of the ano-

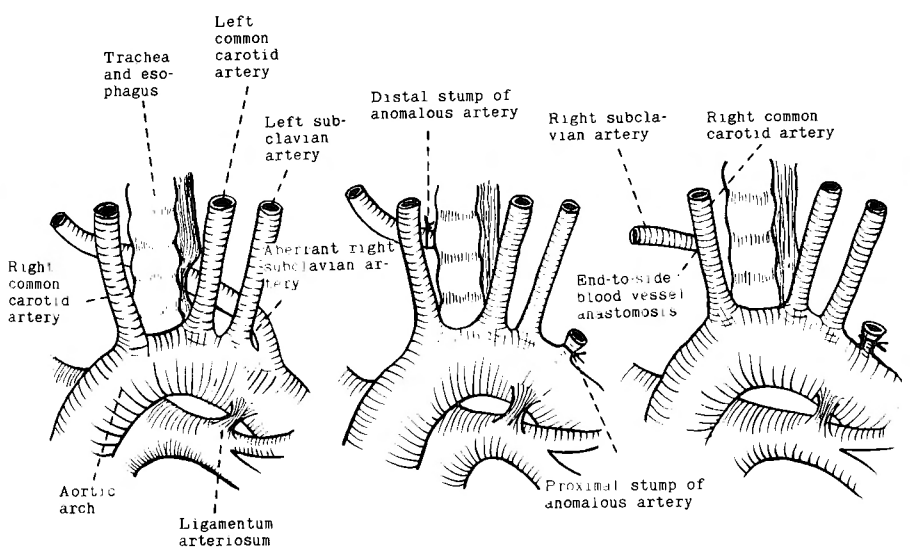

Fig. 21 (a) Anatomical appearance of a case of dysphagia lusoria with aberrant right subclavian artery.
(b) Appearance after the performance of conventional method of operation.
(c) Appearance after the performance of our new method of operation.

malous artery prior to anastomosis resulted in the division of the right internal mammary artery, which nourished the sternoclavicular joint region, the necrosis of this region was induced and thus had prolonged and complicated the postoperative course. Based upon this experience, the operation on our second case was performed at first through the left thoracotomy, as described above, and the anomalous artery was dissected, doubly ligated and severed close to its origin. Then the patient was placed on her back. As second step in the operation, another incision was made in the right cervical region, as described above, and the end-to-side anastomosis was performed between the distal stump of the anomalous artery and the right common carotid artery. These surgical procedures could be performed without any difficulties.

The occlusion test of the right common carotid artery by MATAS, prior to operation, can reveal the possible development of harmful side-effects caused by its occlusion at the time of blood vessel anastomosis. According to Prof. INOKUCHI's experience in 14 cases of common carotid artery replacement, the permissible time of its occlusion ranged from 10 to 50 minutes (mean values 14-15 minutes).[12] In our cases, the blood flow in the right common carotid artery was interrupted for about 25 minutes at the time of anastomosis, but no harmful side-effect was observed. It is possible to shorten the occlusion time and to make the operation safer by the use of blood vessel suturing apparatus. On the other hand, the operator should not confuse the dilated first portion of the anomalous artery with the posterior limb of the double aortic arch, and he must pay attention to the fact that there may be a failure of right recurrent nerve, the right vertebral artery originating in the right common carotid artery etc., in such cases. The operator also has to confirm the true nature of the anomalous artery prior to its ligation and division, by feeling the pulse of radial and common carotid arteries after the temporary interruption of their blood flow. Great emphasis must be laid upon the fact that constriction of the esophagus is not caused solely by the vessel, but that the constriction is likewise produced by fibrous bands or sheaths which accompany this vessel. Failure to cut these strands or bands will give only partial release of an incarceration of the esophagus. It is also important to recognize the presence of a retroesophageal subclavian artery at the time of operation for pulmonic stenosis, and to appreciate its value in using it to create an artificial ductus arteriosus.[3] Before anastomosis to the pulmonary artery the anomalous artery may be brought anterior to the right bronchus and esophagus, if it is necessary.

The right radial pulse was a little feeble for about six hours following the operation. It was thought to be due to the temporary arterial spasm caused by the removal of adventitia at the time of anastomosis, as seen after the performance of peri-arterial sympathectomy by LERICHE.[13]

SUMMARY AND CONCLUSION

Recently we encountered two cases of dysphagia lusoria due to the aberrant right subclavian artery. These cases were successfully operated upon by a new approach, in which the anomalous artery was doubly ligated and divided close to its origin and the end-to-side anastomosis was performed in the cervical region, between its distal stump and the right common carotid artery. Since GROSS carried out the first operation on a four-

month-old baby with this anomaly on December 26, 1945, a rather simple method of operation has been prevailing, in which the anomalous artery is only ligated and divided close to its origin. This method may be sufficient for the patients varying in age from three weeks to six years. However, more and more, this anomaly affecting the over-40-year-old persons is probably coming to light nowadays, because of the increasing utilization of angiocardiography as a diagnostic or prognostic measure in patients with thoracic disease. For such cases, our new method of operation, using the blood vessel anastomosis, should definitely be adopted.

The gist of the present article was reported by KOICHI ISHIGAMI before the 15th General Meeting of the Japanese Association for Thoracic Surgery on October 31, 1962.

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和文抄録

異型右鎖骨下動脈によるDysphagia lusoriaに対する新手術法

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Dysphagia lusoriaとは，1789年Bayfordによつて案出された名称であつて，大動脈およびその分枝の異常によつて惹起される嚥下困難を初めとする症候群をいう．最も多い型式は，異型右鎖骨下動脈（右鎖骨下動脈が正常の場合のように腕頭動脈から起始せずに，大動脈弓の最終分枝として下行大動脈起始部背面から起始し，通常は食道と脊柱との間を走つて右胸尖部に達している場合），重複大動脈弓および左側動脈靱帯を伴う右側大動脈弓である．われわれは最近不定の，次第に増強する嚥下困難を訴えた65才女子および67才女子における異型右鎖骨下動脈の症例に対して，異常動脈をその大動脈起始部に近く結紮，切断し，末梢側切断端を右総頸動脈に用手的に端側吻合することによつて治癒せしめえた．これらの症例は本邦最初の手術治験例である．

本症は1945年11月26日，Gross によつて初めて手術に成功されて以来，その治療対象がほとんど専ら6才以下の幼年者に限られていたため，結紮，切断という単純な外科手術で十分とされて来たが，その発生頻度（0.2～1.4%）から考えて，今後40才以上の中年以降の症例の発見率が増加するものと考えられる．このような異常動脈の硬化性変化や動脈瘤様変化に基ずく40才以上の有症状例に対しては，われわれの提唱する，血管吻合を応用した新手術法を採用すべきである．