ANAESTHESIA FOR SURGICAL CORRECTION OF VASCULAR RING*

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ANOMALIES of the arch of the aorta and the vessels arising from it are not uncommon. In themselves they do not endanger life, nor are they usually associated with congenital heart disease. When they cause obstruction of the oesophagus or trachea, however, surgical intervention is often needed to prevent an early respiratory death.

HISTORY

One of the earliest references in the literature seems to be that of Hommel in 1737, whose case prompted Quain¹ to describe these anomalies in detail in 1844. The obstruction of the oesophagus associated with an aberrant right subclavian artery was graphically described by Bayford² in 1794, whose patient "worn out with fatigue and famine . . . sank into the grave" at the age of 62 years. Gross³ reported the first successful operation to break a vascular ring in 1945. Ten years later⁴ he reviewed experiences in 70 cases with five deaths, all in the type known as double aortic arch. Potts⁵ in 1956 described his experiences with 28 cases, six of whom died. According to Apley,⁶ five clinical reports of these anomalies had appeared in the British literature by 1957. Neuhauser⁷ has been prominent in focussing attention on the radiological diagnosis. Many centres doing paediatric surgery have now had experience with these cases.

CLASSIFICATION

An excellent account of the embryology and a comprehensive classification is given by Harley.⁸ Many of the possible variants he lists have yet to be described. In addition, Harley gives a useful clinical classification into three groups:

(1) Those vessels which compress only or mainly the oesophagus. This comprises mainly the subclavian artery arising from the distal part of the aortic arch and going behind the oesophagus to the opposite axilla. Also included here is a ductus arteriosus communicating with a right-sided aorta and passing behind the oesophagus to the left pulmonary artery (Fig. 1).

(2) Vessels compressing only the trachea such as anomalous innominate or left common carotid arteries (Fig. 3).

(3) Those vessels which compress both structures. This includes all other variants of vascular ring, such as double aortic arch (Fig. 2).

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FIGURE 1

FIGURE 2

FIGURE 3

CLINICAL PICTURE AND DIAGNOSIS

Not all vascular ring anomalies give rise to serious symptoms. When these occur the initial picture depends on whether the oesophagus or trachea is more compressed. Oesophageal obstruction in infancy leads to regurgitation and choking during and after feeding, and inevitably causes aspiration pneumonia. When tracheal compression is present the infant has continuous difficulty in breathing with obstructive sounds which can be heard both in inspiration and expiration. In severe cases the baby assumes a hyper-extended position to make breathing easier. Attacks of cyanosis and unconsciousness occur. Respiratory infections and broncho-pneumonia are common and prolonged. With these complications the signs of respiratory obstruction are very severe. Tracheostomy may seem indicated but is usually useless, for this tracheal obstruction is often just above the carina. One of our patients (Case 2) had a tracheostomy to tide him over a severe bout of pneumonia, but his obstruction was subsequently found to be mainly oesophageal.

Radiological examination of the oesophagus and trachea using contrast media in very small amounts usually makes the diagnosis with amazing accuracy. According to Gross⁴ radiological evidence of oesophageal and tracheal compression in infancy in the absence of a mediastinal tumor is almost diagnostic of some form of vascular ring. The oesophagus is usually seen to be displaced forward and indented posteriorly at about the level of thoracic 2 or 3. The degree and site of tracheal compression are usually reported accurately by the radiologist. This compression will be seen to vary, sometimes considerably, with the phases of respiration. Time is well spent by the anaesthetist in study of these films. We have not found tracheoscopy and oesophagoscopy necessary, nor is angio-cardiography usually indicated. These infants are often very ill.

CASE HISTORIES AND RESULTS

Some of the main points concerning the twelve patients in this series are shown in Table I. Surgical correction of the vascular anomalies was performed in eight of these patients without mortality and with satisfactory clinical and radiological

TABLE I
CASES WITH VASCULAR RING ANOMALIES

Case	Age	Predominant symptoms	Type of lesion	Operation	Anaesthetic difficulties	Postoperative complications	Result
1	20 mos.	Difficulty in swallowing, respiratory infections	Right descending aorta, aberrant left subclavian, and ligamentum arteriosus	Division left subclavian and ligamentum arteriosus	No airway problems	None	Good; alive and well
2	6 wks.	Severe pneumonia	Aberrant right subclavian artery	Division right subclavian and forward fixation of great vessels	No airway problems	None	Good; alive and well
3	6 mos.	Severe pneumonia	Fallot's Tetralogy and aberrant subclavian artery	None			Still not operated; condition no worse
4	27 mos.	Persistent cough, extensive right-sided pneumonia	Right descending aorta, left ligamentum arteriosum	Division ligamentum arteriosum, forward fixation of great vessels	Momentary obstruction during dissection, around trachea	None	Satisfactory; alive and well
5	6 wks.	Attacks of respiratory obstruction and cyanosis	Double aortic arch with ductus arteriosus	Division anterior arch and ductus arteriosus	Deliberate right lung anaesthesia only; recurrent laryngeal and probably left phrenic nerve also inadvertently cut	Obstruction; bronchoscopy, tracheostomy	Now two years of age; alive and well
6	3 mos.	Blue spells while feeding, severe broncho-pneumonia and status epilepticus	Aberrant right subclavian artery	None			Not operated; died from intercurrent medical disease

TABLE I (Concluded)

Case	Age	Predominant symptoms	Type of lesion	Operation	Anaesthetic difficulties	Postoperative complications	Results
7	2 mos.	Some choking and cyanotic attacks	Probably right aortic arch with aberrant left subclavian artery	none			Has done well without operation
8	4 mos.	Wheezing	Double aortic arch	Division of smaller posterior arch	Intubated beyond obstruction; no difficulties	Left-side pneumonia 4 days postoperatively	Good; alive and well
9	2 wks.	Wheezing, crowing when feeding, and cyanosis	Double aortic arch, right descending aorta, aberrant left subclavian artery, and ductus arteriosus	Division anterior arch and ductus arteriosus	Endotracheal tube placed distal to obstruction; no difficulties	None	Good, clinically and radiologically
10	7 mos.	Repeated attacks of pulmonary infection	Right descending aorta, aberrant left subclavian artery, and ductus arteriosus	Division left subclavian and ductus arteriosus, forward fixation of great vessels	During dissection intermittent obstruction of left main bronchus occurred	None	Has done well
11	1 day	Oesophageal atresia and tracheo-oesophageal fistula	Aberrant right subclavian artery seen during surgery	Repair oesophageal atresia and tracheo-oesophageal fistula; no vascular surgery	None	None	Doing well; no clinical or radiological evidence to justify further surgery
12	1 mo.	Pneumonia	Right-sided arch, right descending aorta, aberrant left subclavian artery, and ductus arteriosus	Division left subclavian and ductus arteriosus, forward fixation of pulmonary artery	None with controlled respiration	No immediate complications; admitted again with aspiration pneumonia	Now alive and well

results. In one of the remaining four patients (Case 11) an aberrant right subclavian artery was seen during surgery done to repair an oesophageal atresia and tracheo-oesophageal fistula. This vessel was not resected and there has been no subsequent indication to do so. In Case 3 an aberrant subclavian artery was diagnosed radiologically in conjunction with a Fallot's tetralogy. This patient awaits surgery for his heart condition. One patient (Case 7) has not had severe enough symptoms to warrant surgery, while the remaining patient (Case 6) died in status epilepticus with overwhelming broncho-pneumonia before surgery could be performed. A more detailed description of the salient points in these case histories follows.

Case 1

A girl was admitted at 20 months of age with a history of frequent respiratory infections and difficulty in swallowing. Radiological examination of the chest revealed that the oesophagus was displaced sharply to the left at the level of thoracic 2 and 3 and that a posterior filling defect was present (Fig. 4). A tracheogram was done under general anaesthesia with nitrous oxide, oxygen, and trichlorethylene. This revealed a minor defect on the right side without any marked narrowing of the trachea. At operation the surgeon found a right-sided descending aorta with the left subclavian artery and the ligamentum arteriosum both arising from it and passing behind the oesophagus (Fig. 5). Both these structures were divided and the postoperative condition was much improved. For anaesthesia nitrous oxide, oxygen, d-tubocurarine, and minimal ether were given. There were no airway problems involved here.

Case 2

This patient was admitted at six weeks of age with very severe pneumonia and respiratory obstruction. The diagnosis of vascular ring was entertained at that time, but because of the respiratory obstruction it was decided to perform tracheostomy. When the patient had sufficiently recovered and the tracheostomy had been removed, a barium swallow revealed that the oesophagus was displaced forward and indented posteriorly at the level of thoracic 2 and 3 (Fig. 6). A tracheogram revealed only a minor defect on the right side of the trachea about 2 cm. proximal to the carina. Anaesthesia for thoracotomy was nitrous oxide, oxygen, and ether by oral endotracheal tube and no airway difficulties were encountered. A right subclavian artery arising from the descending aorta was found coursing behind the oesophagus (Fig. 7). This was divided and the great vessels were freed forward from the front of the trachea and fixed anteriorly to the back of the sternum to prevent compression of the trachea. The child has done well since.

Case 3

This child was admitted at six months of age with severe pneumonia and cyanotic heart disease. Radiological examination of the chest revealed a picture suggestive of Fallot's tetralogy, and barium swallow showed an oblique defect in the oesophagus probably due to an aberrant subclavian artery. He improved on being treated with high humidity and has since done well, not requiring further hospital admissions. He is now three years of age.

Case 4

This 27-month-old child was first admitted with extensive right-sided pneumonia. This cleared up with antibiotic therapy except for a persistent cough. About four and one-half months later she was re-admitted when a barium swallow revealed that the oesophagus was compressed both posteriorly and bilaterally just above the level of the carina. Slight narrowing of the trachea was evident at the same level. For thoracotomy, anaesthesia was induced with cyclopropane following rectal pentothal; succinylcholine was used for

FIGURE 4

intubation; and respirations were controlled with the Jefferson Ventilator, Gallamine being given for longer acting muscle relaxation. After a fairly lengthy surgical dissection a ligamentum arteriosum, communicating with the left pulmonary artery and a right-sided descending aorta, was found and divided. As recommended by Gross, the great vessels were also fixed forward to the sternal region by sutures. Since there was no great compression of the trachea, the airway was quite clear during the early part of anaesthesia. A monaural stethoscope placed at the back of the chest enabled the anaesthetist to monitor the breath sounds continuously. However, during the dissection

FIGURE 5

FIGURE 6

anterior to the trachea the respiratory exchange became inadequate. The surgeon stopped work on request, the endotracheal tube was adjusted, and anaesthesia was manually controlled from this point onward. Postoperatively the child did well, apart from a slight cough, and has been free of symptoms since.

Case 5

This little boy was admitted at six weeks of age with a history of respiratory difficulty from birth with severe attacks of respiratory obstruction and cyanosis. Radiological examination of the chest revealed a persistent indentation of the oesophagus posteriorly

FIGURE 7

and on the right and left just above the level of the carina together with considerable narrowing of the trachea at the same level (Fig. 8). At thoracotomy the patient was found to have a double aortic arch and ductus arteriosus. The larger arch (developmentally the right arch) passed posteriorly to the oesophagus as usual, but the smaller arch (anterior) gave off the left common carotid and left subclavian arteries. Accordingly, this arch was divided between the left common carotid and subclavian arteries. It is important to leave the left common carotid artery coming off the proximal part of aortic arch rather than have the blood enter it in a retrograde fashion. The ductus arteriosus was divided and the vessels freed forward off the front of the trachea and fixed to the back of the sternum. Unfortunately, during the dissection, the recurrent laryngeal

FIGURE 8

nerve and probably the left phrenic nerves were cut—this led to trouble later. Anaesthesia was induced with cyclopropane. An endotracheal tube was inserted past the obstruction and throughout most of the procedure only the right lung was ventilated. Controlled respiration using small doses of succinylcholine intermittently was employed. The patient did well for some time after extubation but then suddenly became obstructed and emergency bronchoscopy and tracheostomy were needed. Bronchoscopy was done on two other occasions during a stormy convalescence. He was finally discharged two and one half months after operation. He is now two years of age and doing well.

Case 6

This infant was admitted at three months of age with a history of blue spells when feeding. She only weighed 7 lb at this time. She was discharged and readmitted at seven months of age in status epilepticus and with severe broncho pneumonia from which she subsequently died. At autopsy an aberrant right subclavian artery was found.

Case 7

This little boy has had a long history of admissions to hospital from two months of age with attacks of choking and cyanosis on crying. Radiological examination of the oesophagus showed posterior indentation of the oesophagus at the level of thoracic 2 and 3 and bilateral constriction. The trachea did not seem to be much narrowed. The most recent tracheograms and oesophagrams have suggested a right sided aortic arch. There is not enough evidence of obstruction of the trachea or oesophagus to justify surgical intervention.

Case 8

At four months of age this boy was admitted with a history of wheezing. Before admission a diagnosis of an enlarged thymus was made at another hospital and treated by radiation. As this did not improve the complaint the patient was admitted to our hospital. Radiological examination of the chest revealed that the oesophagus and trachea were both compressed and narrowed, the indentation in the trachea being approximately 2 cm proximal to the carina (Figs 9 and 10). As in many of these cases the narrowing of the trachea was seen to vary considerably with different phases of respiration. The diagnosis was vascular ring and thoracotomy was accordingly performed. For this the

FIGURE 9

FIGURE 10

patient was induced with cyclopropane following a small dose of rectal pentothal, intubated following succinylcholine relaxation and maintained on nitrous oxide, oxygen, and ether. A No. 1 Magill endotracheal tube was used and there was no great trouble with the airway, to our surprise. A double aortic arch was found with the smaller one going posteriorly. This was divided and the constriction relieved (Fig. 11). The infant had evidence of left-sided broncho pneumonia developing about four days postoperatively. This cleared up and the child was discharged in good condition.

Case 9

This boy was admitted at two weeks of age with a history of wheezing respiration crowing when feeding, and attacks of cyanosis. Visualization of the oesophagus and trachea by contrast medium revealed that the oesophagus was both deviated and compressed at about the level of the aortic arch and that the trachea was indented on the right and posteriorly about 1.3 cm. above the carina (Fig. 12). An effort was made to place the tip of the endotracheal tube distal to the obstruction of the trachea. This was apparently successful and anaesthesia was relatively uneventful. At thoracotomy a double aortic arch with a right-sided descending aorta was dissected out. The anterior limb was divided, as well as a ductus arteriosus, and the trachea and oesophagus freed out (Fig. 13). Postoperatively the chest gradually cleared and re-examination showed that the obstruction to the oesophagus and trachea has been largely removed.

Case 10

This seven-month-old boy had repeated attacks of pulmonary infection especially in the right upper lobe. Bronchoscopy on three occasions was negative. Examination of the oesophagus using contrast medium revealed marked posterior indentation and displacement to the left of this structure at the level of thoracic 3. Narrowing of the trachea just above the carina was also seen. At operation a right-sided descending aorta with the left subclavian coming off and coursing behind the oesophagus and a patent ductus arteriosus completed the ring. After careful dissection the left subclavian and ductus arteriosus were divided. The pulmonary artery and left common carotid were sutured forward to the sternum to release pressure on the trachea. Anaesthesia was induced with cyclopropane and intermittent succinylcholine given to enable respirations to be controlled throughout the procedure. During the dissection it was very difficult at times to inflate the left lung and this was to be expected. A reinforcing dose of 0.2 mg. of

FIGURE 11

FIGURE 12

atropine was given intravenously about three hours after the start of surgery and during the dissection around the oesophagus when some heart slowing was noted. The post-operative course was fairly uneventful and the last X-rays showed the oesophagus not to be obstructed.

Case 11

This infant girl was admitted to our hospital a few hours after birth with a diagnosis of oesophageal atresia and tracheo-oesophageal fistula. This was satisfactorily repaired. During the procedure an aberrant right subclavian artery was noted by the surgeon who judged that it was not compressing the trachea and left it alone. Subsequently this vessel was seen pulsating on oesophagoscopy performed at eight months of age, although it was not causing obstruction. Radiological examination of the chest using a cine machine demonstrated that there is some slight indentation of the oesophagus by the aberrant subclavian artery and some occasional narrowing of the trachea. However, so far there have been no symptoms to justify further surgical intervention.

Case 12

An infant boy was admitted at one month of age because of an upper respiratory infection and right upper lobe pneumonia. Radiological examination of the oesophagus and trachea revealed posterior indentation of the oesophagus and an oblique defect probably due to an aberrant vessel. There was as well a well-defined narrowing of the trachea just above the carina. After induction with cyclopropane and intubation following succinylcholine, a No. 1 Davol endotracheal tube was easily introduced past the obstruction to the right main bronchus. This was confirmed by continuous auscultation of the chest. The tube was then gradually withdrawn till aeration of both lungs was present. Respiration was controlled throughout, and hypothermia to 31° C. induced. Rewarming to 33.0° C. was done before the end of surgery. A right-sided aortic arch and right descending aorta were found with the left subclavian artery and the ductus arteriosus both coming off on the right side and winding round posteriorly to the oesophagus. These vessels were divided and the pulmonary artery fixed anteriorly to the sternum to relieve compression on the trachea. During the dissection, spontaneous

FIGURE 13

respiration inadvertently occurred on one occasion and aeration at once became inadequate. With controlled respiration, on the other hand, ventilation was always efficient. The postoperative course of this infant from a respiratory point of view was fairly satisfactory. The baby was discharged 16 days postoperatively but has had to be readmitted on two occasions because of pneumonia following aspiration. However, he now seems to be doing quite well.

SUGGESTED MANAGEMENT OF ANAESTHETIC PROBLEMS

The preoperative condition of these patients is often serious. They must frequently be accepted for anaesthesia with severe respiratory infection still present. It may be almost impossible to keep a clear airway throughout the surgical procedure. The immediate postoperative period on occasion may be worse than before surgery.

The anaesthetist must carefully assess the patient both from the clinical and radiological findings. The greatest difficulty and stormiest postoperative course may usually be expected in patients with double aortic arch. The site of tracheal obstruction, when present, is important to the anaesthetist. If situated just above the carina, placing the tip of the endotracheal tube distal to the obstruction may mean using one lung anaesthesia only, as in Case 5. A selection of endotracheal tubes of different sizes and lengths must be carefully assembled.

We have favoured the use of fairly heavy premedication with atropine only, usually 0.24 mg. to 0.3 mg. in infants under six months of age. In one of our patients (Case 10) a reinforcing dose of atropine was given because of heart slowing during the dissection around the oesophagus. Hypoxia might have contributed to this bradycardia.

In the larger patients we have occasionally given rectal pentothal to make induction of anaesthesia easier. Usually, however, anaesthesia was induced with cyclopropane and oxygen. Smith⁹ has questioned the advisability of trying to insert the endotracheal tube past the tracheal obstruction, while Wetchler and McQuiston¹⁰ have tended to favour this technique. Wherever the tube is placed, controlled respiration is absolutely essential. The oxygen demands of the muscles of respiration may make a marginal hypoxic state serious. In addition, whereas adequate ventilation can be maintained with controlled respiration, the onset of spontaneous breathing often leads to immediate obstruction from mediastinal movement. Finally, only with controlled respiration can the surgeon have a quiet field for what is usually a lengthy and dangerous dissection. Intermittent succinylcholine together with minimal fluothane using a T-piece technique works well. We have tried to place the tip of the endotracheal tube distal to the tracheal narrowing when this is marked. Moderate hypothermia to 32.0° C. helps to tide the patient over periods of hypoxia which can occur during the dissection. The breath sounds must be continuously monitored by stethoscope, which can be placed in the axilla opposite to the surgical incision or at the back of the chest. The anaesthetist must watch the field, as sudden obstruction is usually related to surgical interference. The surgeon and his assistants must immediately desist when this occurs. Rapid blood loss can occur and must be immediately replaced. The infant's blood-pressure cuff and monaural stethoscope are very important pieces of paediatric equipment here.

We have not resorted to cutting side-holes in the endotracheal tube, although a single suitably-placed hole for the upper lobe bronchus may help when the right main bronchus is deliberately intubated. Neither have we encountered any case where the lumen of the trachea was so small that a conventional endotracheal tube of suitable size would not pass. This difficulty has been described by Wetchler and McQuiston. With the range of small plastic endotracheal tubes now commercially available even the smallest trachea can be intubated. Frequent suctioning through the endotracheal tube must be done regularly, whether the breath sounds are moist or not. Almost all the dissection is around the carina* and the trauma involved frequently causes bleeding into the lumen of the trachea.

In the postoperative period the patient is placed in an atmosphere of maximal humidity. Antibiotics and steroids may be necessary. Tracheostomy is to be avoided although sometimes, as in Case 5, it may be unavoidable; in this case cutting the recurrent laryngeal nerve complicates matters. An endotracheal tube, small enough to pass through the tracheostomy tube and beyond the tracheal obstruction must be to hand. Moderate hypothermia, under medical supervision, in the postoperative period may be needed to tide the infant over recurring bouts of obstruction and hypoxia. These patients ought to have follow-up radiological examinations some months after operation to assess the effectiveness of surgery.

SUMMARY

The literature on vascular ring anomalies is briefly reviewed. The results of some recent cases at The Children's Hospital, Winnipeg, are presented.

The anaesthetist must study the clinical and radiological findings carefully. Controlled respiration through an endotracheal tube of suitable size and length is used together with moderate hypothermia. The airway is precarious and complete co-operation between anaesthetist and surgeon is essential. Blood loss may be sudden and severe.

In the postoperative period, tracheostomy is to be avoided if possible. Maximum humidity, antibiotics, and steroids are needed.

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RÉSUMÉ

Une grande partie de l'anatomie des anomalies populairement connues sous le nom d'anneau vasculaire, a été démontrée depuis plus de deux siècles, mais c'est Grosse de Boston qui a opéré le premier cas avec succès en 1945. Depuis cette époque de nombreux centres contribuent des rapports qui ont permis de connaître davantage les problèmes à la fois en chirurgie et en anaesthésie.

Cet article est un rapport d'expériences avec des cas semblables, opérés à l'Hôpital des Enfants, de Winnipeg, dans les années récentes.

Les anomalies vasculaires peuvent être divisées cliniquement en trois catégories. Dans la première catégorie, le vaisseau anormal ou les vaisseaux anormaux resserrent surtout l'oesophage; dans la seconde, c'est la trachée-artère qui est obstruée; mais dans la troisième classe un anneau vasculaire complet est la cause de l'obstruction à la fois de l'oesophage et de la trachée-artère.

C'est souvent dans un état sérieux que ces malades sont acceptés pour l'anaesthésie, aussi ils souffrent parfois d'infections respiratoires sévères. Il est obligatoire de contrôler le souffle pendant toute la procédure, et le risque d'anoxémie qui très souvent arrive causé par l'obstruction, est diminué par l'usage d'une hypothermie modérée à 32° ou 31° C.

Dans un cas sévère nous avons essayé de placer notre tube après l'obstruction dans la trachée-artère. En employant un stéthoscope bien placé, les sons de la respiration doivent être continuellement auscultés. Le traumatisme chirurgical bien souvent est la cause de saignement dans la trachée, si bien qu'il est nécessaire d'aspirer le tube à intervalles réguliers et fréquents. L'anaesthésiste doit regarder la procédure chirurgicale tout le temps, car un saignement soudain pourrait arriver. Si le chirurgien est la cause de l'obstruction trachéale, on lui demande d'arrêter immédiatement.

Après l'opération il vaut mieux éviter la trachéotomie. Un maximum d'humidité, des antibiotiques et de cortisone sont administrés. Une hypothermie modérée aurait peut-être besoin d'être continuée pendant quelques jours.

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