Double Aortic Arch Surgery

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Abstract

Double aortic arch is a congenital anomaly that occurs due to abnormality in development of arches during foetal life. Symptoms occur secondary to compression of adjacent structures by the arches. Surgery remains the definitive treatment for this pathology.

Introduction

Aortic arch anomalies where first described in the 17th century. There are various types of this anomaly. Double aortic arch was described by Hommel in 1763.[1] The first surgical procedure for this condition was done in 1945 by Dr Gross who did division and ligation of vascular ring to relieve oesophageal obstruction.[2]

Prevalence of this anomaly is 0.5 to 1% of all the congenital cardiovascular abnormalities. It usually presents in neonatal and infantile age group with symptoms of recurrent respiratory tract infection and regurgitation.[3] If untreated the symptoms progessively worsen and stridor can develop due to tracheomalacia. Double aortic arch has three subtypes

1. Right dominant arch(75%).
2. Left dominant arch(20%).
3. Codominant arches(5%)

Diagnosis is based on demonstration of a vascular ring encircling and compressing the trachea or oesophagus with the help of CT thorax with contrast.[4]

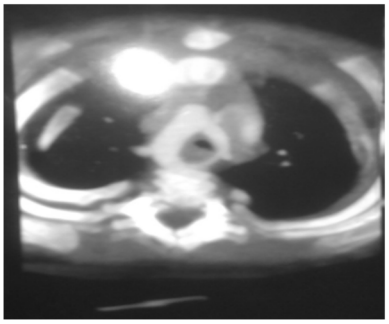
Surgery remains the definitive treatment for this anomaly. It should be performed at the earliest before the child develops complications due to regurgitation and tracheomalacia.

The idea behind this case report presentation is to highlight the fact that though arch anomalies are rare, surgery is a simple and definitive treatment option.

Case Report

3 months 4.5 kg male child, presented with chief complaints of noisy breathing, difficulty in swallowing along with regurgitation, persistent dry cough, and breathlessness increased after feeding. Parents also gave history of recurrent respiratory tract infection with history of hospitalisation once at the age of 2 months

During this hospitalisation he was investigated. His blood investigation were normal, chest X-ray did not show any cardiomegaly or pneumonic patch that could correlate with his symptoms. 2D ECHO also did not show any congenital cardiac abnormality. Therefore CECT scan was done which showed double aortic arch compressing the trachea and oesophagus.



He was referred to our hospital for further

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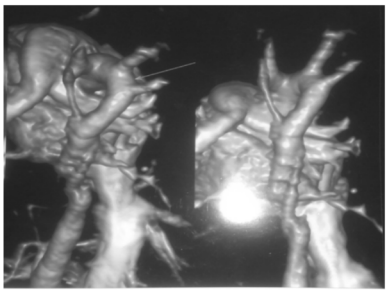
management of this anomaly.

On examination,

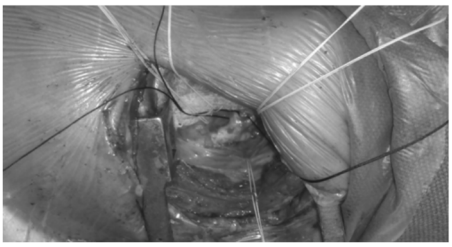
- Cardiovascular system: no audible murmurs
- Respiratory system: bilateral conducted sounds present.
- P/A examination: not contributory.

Investigation

- X-ray chest: did not show any obvious abnormality.
- Electrocardiogram: no significant changes seen.
- Echocardiogram: showed 2 mm pfo and patent ductus arteriosus.
- CT aortogram with 3d reconstruction: double aortic arch compressing the trachea and oesophagus was seen. Right aortic arch was dominant and major arch, measuring 5.6 mm in diameter. Left aortic arch was nondominant and minor arch measuring 4.0 mm in diameter. Left common carotid and left subclavian artery arises from left minor arch while right common carotid and right subclavian artery arises from right major arch. Complete vascular ring was seen around the trachea.



Surgery was planned. Right radial and left femoral arterial lines were taken and saturation probes was placed on left hand and right foot . Under general anaesthesia, right lateral position was given and a lateral thoracotomy was done in the 4th intercostal space. Medisternal pleura was reflected from the aorta and stay sutures taken. Double aortic arch along with the patent ductus arteriosus were defined.



Findings: right and left aortic arches along with the PDA formed a vascular ring that compressed the trachea and oesophagus.

Right and left aortic arch and patent ductus arteriosus confirmed by temporarily clamping each vessel and seeing for change in saturation and arterial pressure tracing. With this left aortic arch was confirmed as nondominant

PDA was ligated by 2-0 silk, clipped and divided. Left aortic arch was ligated with 2-0 silk, beyond the left subclavian artery and then divided. Cut ends were oversewn by prolene 6-0 and was fixed to the prevertebral fascia. Haemostasis was confirmed. Left pleural drain was placed and thoracotomy was closed in layers.

Patient extubated on next day morning. Postoperative recovery was uneventful. All the presenting symptoms were relieved.

Discussion

Being a rare anomaly, diagnosis of double aortic arch solely based on clinical presentation is difficult. It can be considered as one of the causes for the respiratory and regurgitation symptoms only after all the common aetiologies have been ruled out. Confirmation of the diagnosis is based on CT scan and CT aortogram is the gold standard for this as it helps to confirm the diagnosis, classify the type of anomaly, defines the dominant arch with anatomical orientation of branches of arch, help to plan the surgical procedure and follow up the patient post procedure for any complication.[4]

Ideally fibreoptic bronchoscopy should be done in all patients to detect tracheomalacia and grade its severity.[5] However due to the age, size of patient it was not done.

Clamp test is important intraoperatively in order to define the dominant and nondominant arch.[6] Surgical division and ligation of

nondominant arch removes the compression on trachea and oesophagus and relieves all the symptoms. Patent ductus arteriosus should be looked for and it should also be divided and ligated to prevent any residual compression. Cut ends should be plastered to prevertebral fascia to prevent the rare possibility recannalisation.[7]

Extubation should be delayed and arrangement for reintubation should be present due to the risk of development of stridor secondary to tracheomalacia. Hence airway management is important in this set of patients.[8]

Conclusion

With proper understanding of aortic arch anatomy, double aortic arch surgery becomes simple and safe. Also surgical treatment is a definitive option with good outcome and excellent long term prognosis. Hence once detected surgery should be encouraged and performed at the earliest.

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Can healthy people gain from health apps?

Many health apps are aimed at people with no diagnosis: for example, apps that allow users to track their calorie intake and exercise.

But another technological wave is rising: medical apps for smartphones and tablets. These supposedly promote mental health, and sleep, cause weight loss, control food allergy, aid self diagnosis, manage pain and help in every other conceivable medical condition. Indeed, some are endorsed by the NHS.

These tens of thousands of health apps are perhaps mostly harmless (and likely useless).

When used alongside wearable gizmos, apps offer continuous physical monitoring of things such as foetal heartbeat in pregnancy, blood pressure, heart rate, and even pulse oximetry.

So what will users of these apps discover? How common brief arrhythmias are in the normal population? How often our blood pressure might be high? How widely normal oxygen saturations can vary?

The truth is that these apps and devices are untested and unscientific, and they will open the door of uncertainty. Make no mistake: diagnostic uncertainty ignites extreme anxiety in people. We must reflect on what we might lose here, rather than what we might gain. Will apps simply empower patients to overdiagnosis and anxiety?

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