Anomalous subaortic position of the brachiocephalic (innominate) vein: a review of published reports and report of three new cases

LEON M GERLIS, SIEW YEN HO

From the Department of Paediatrics, National Heart and Lung Institute, London

SUMMARY Anomalous courses of the left innominate vein have rarely been described in anatomical specimens. Investigative techniques such as angiography and echocardiography have brought to light more instances of this anomaly. Three more cases identified by anatomical study are described. Earlier cases were reviewed to assess the type of associated cardiac malformations. Clinically, the abnormality is regarded as benign. When it is recognised during investigation it should alert the clinician to the possibility of associated malformations. Features commonly seen in tetralogy of Fallot—right aortic arch, ventricular septal defect, and right ventricular outflow obstruction—were common in patients with anomalous subaortic innominate veins.

The left brachiocephalic (or innominate) vein extends from the junction of the left internal jugular and left subclavian veins to the junction of the right brachiocephalic (or innominate) vein and the (right) superior caval vein. The normal course is obliquely downwards and to the right, passing in front of the left subclavian, left common carotid, and brachiocephalic arteries, beneath the aortic arch. Very rarely, this vein (or its analogue) follows an anomalous course, passing from left to right below the arch of the aorta, to enter the (right) superior caval vein below the orifice of theazygos vein. Kershner first described this anomaly a hundred years ago, and we are aware of at least another 21 cases (table). We report three further cases.

Although the malformation in itself seems to be of no functional importance, we assessed its importance in terms of associated conditions and its possible relevance to subsequent operations.

Case reports

CASE 1

This girl died when she was 5 years 11 months old after cerebral ischaemia after attempted surgical correction of tetralogy of Fallot. Postmortem examination of the heart showed usual (solitus) atrial arrangement with concordant atrioventricular and ventriculoarterial connections, infundibular and valvar pulmonary stenosis, subaortic ventricular septal defect, and several large aortopulmonary anastomoses. There was a right sided aortic arch. The site of the arterial duct was not apparent. The large left brachiocephalic vein passed behind the ascending aorta and below the aortic arch before entering the superior caval vein (vena cava), below the level of theazygos vein and just above its junction with the right atrium (fig 1).

CASE 2

This male infant died of complex cardiac malformation at one day. There was the usual (solitus) atrial arrangement with concordant atroventricular connection and double outlet right ventricle with a dextroposed aortic orifice overriding a perimembranous ventricular septal defect. There appeared to be an ectopic portion of atroventricular valve, complete with leaflet tissue, tendinous cords, and papillary muscles, situated between the trabecular and infundibular components of the right ventricle. The unguarded pulmonary artery orifice showed no trace of arterial valve leaflets. The arterial duct was short and widely patent. The aortic arch was left sided and had normal branches. The left internal jugular vein continued into a descending vein, which passed under the aortic arch from left to right, anterior to the arterial duct and between the arch and the pulmonary trunk. It then ran behind the ascending aorta to enter the lower portion of the superior
Anomalous subaortic position of the brachiocephalic (innominate) vein

Table  Reported cases of subaortic left brachiocephalic vein

<table>
<thead>
<tr>
<th>Case</th>
<th>Authors</th>
<th>Year</th>
<th>Reference No</th>
<th>Age</th>
<th>Sex</th>
<th>Diagnosis</th>
<th>Relation to arterial duct</th>
<th>Aortic arch</th>
<th>Other cardiovascular malformations</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Kerschner</td>
<td>1888</td>
<td>2</td>
<td>?(child)</td>
<td>?</td>
<td>Necropsy</td>
<td>?</td>
<td>L</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>Daser</td>
<td>1902</td>
<td>6</td>
<td>66 yr</td>
<td>M</td>
<td>Necropsy</td>
<td>Anterior</td>
<td>L</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>Ghon</td>
<td>1908</td>
<td>7</td>
<td>4½ mo</td>
<td>F</td>
<td>Necropsy</td>
<td>Posterior</td>
<td>R</td>
<td>Absent L 4th arch, isolated L subclavian artery</td>
</tr>
<tr>
<td>4</td>
<td>Nützel</td>
<td>1914</td>
<td>8</td>
<td>74 yr</td>
<td>M</td>
<td>Necropsy</td>
<td>?</td>
<td>L</td>
<td>Right superior pulmonary vein to superior caval vein</td>
</tr>
<tr>
<td>5</td>
<td>Martin</td>
<td>1931</td>
<td>9</td>
<td>17 yr</td>
<td>M</td>
<td>Necropsy</td>
<td>Anterior</td>
<td>R</td>
<td>Fallot</td>
</tr>
<tr>
<td>6</td>
<td>Walter</td>
<td>1931</td>
<td>10</td>
<td>19 yr</td>
<td>M</td>
<td>Necropsy</td>
<td>Anterior</td>
<td>L</td>
<td>Subclavian venous anastomosis</td>
</tr>
<tr>
<td>7</td>
<td>Adachi</td>
<td>1933</td>
<td>11</td>
<td>20 yr</td>
<td>M</td>
<td>Necropsy</td>
<td>Anterior</td>
<td>L</td>
<td>None</td>
</tr>
<tr>
<td>8</td>
<td>Adachi</td>
<td>1933</td>
<td>11</td>
<td>41 yr</td>
<td>M</td>
<td>Necropsy</td>
<td>Posterior</td>
<td>L</td>
<td>None</td>
</tr>
<tr>
<td>9</td>
<td>Friedman</td>
<td>1945</td>
<td>12</td>
<td>66 yr</td>
<td>M</td>
<td>Necropsy</td>
<td>Anterior</td>
<td>L</td>
<td>L jugular vein anomaly</td>
</tr>
<tr>
<td>10</td>
<td>Roberts et al</td>
<td>1951</td>
<td>1</td>
<td>7 yr</td>
<td>F</td>
<td>Angio</td>
<td>Subpulmon art</td>
<td>L</td>
<td>None</td>
</tr>
<tr>
<td>11</td>
<td>Sherman</td>
<td>1963</td>
<td>4</td>
<td>?</td>
<td>?</td>
<td>Necropsy</td>
<td>Anterior</td>
<td>L</td>
<td>Pulmonary ateria VSD</td>
</tr>
<tr>
<td>12</td>
<td>Sherman</td>
<td>1963</td>
<td>4</td>
<td>?</td>
<td>?</td>
<td>Necropsy</td>
<td>Anterior</td>
<td>L</td>
<td>Pulmonary ateria VSD</td>
</tr>
<tr>
<td>13</td>
<td>Yoshida et al</td>
<td>1975</td>
<td>13</td>
<td>68 yr</td>
<td>M</td>
<td>Necropsy</td>
<td>Anterior</td>
<td>L</td>
<td>None</td>
</tr>
<tr>
<td>14</td>
<td>Kitamura et al</td>
<td>1981</td>
<td>14</td>
<td>69 yr</td>
<td>M</td>
<td>Necropsy</td>
<td>Anterior</td>
<td>L</td>
<td>None</td>
</tr>
<tr>
<td>15</td>
<td>Cloez et al</td>
<td>1982</td>
<td>15</td>
<td>1½ yr</td>
<td>M</td>
<td>Echo and angio ?</td>
<td>L</td>
<td>Fallot</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>Smallhorn et al</td>
<td>1985</td>
<td>5</td>
<td>?</td>
<td>?</td>
<td>Echo ?</td>
<td>R</td>
<td>Central pulmonary arteries absent in one</td>
<td></td>
</tr>
<tr>
<td>23</td>
<td>Gerlis et al</td>
<td>1988</td>
<td>—</td>
<td>5 yr 11 mth</td>
<td>F</td>
<td>Necropsy</td>
<td>No duct</td>
<td>R</td>
<td>Fallot</td>
</tr>
<tr>
<td>24</td>
<td>Gerlis et al</td>
<td>1988</td>
<td>—</td>
<td>1 day</td>
<td>M</td>
<td>Necropsy</td>
<td>Anterior</td>
<td>L</td>
<td>Complex</td>
</tr>
<tr>
<td>25</td>
<td>Gerlis et al</td>
<td>1988</td>
<td>—</td>
<td>7 wk</td>
<td>F</td>
<td>Necropsy</td>
<td>No duct</td>
<td>R</td>
<td>Common arterial trunk</td>
</tr>
</tbody>
</table>

Angio, angiography; echo, echocardiography; subpulmon art, subpulmonary artery; VSD, ventricular septal defect.

Fig 1 Photograph and diagram of case 1 showing the anomalous left brachiocephalic vein (ALBV) running behind the ascending aorta (Ao) and below the right aortic arch (RAA) before entering the superior caval vein (SCV).

Fig 2 Photograph and diagram of case 2 showing the anomalous left brachiocephalic vein (ALBV) passing below the aorta (Ao). AD, arterial duct; PT, pulmonary trunk.

caval vein between the entry of the azygos vein and the junction with the right atrium. Because the specimen was damaged it was impossible to ascertain the connection between this anomalous vein and the left subclavian vein. There was, however, no evidence of any other route of venous drainage from the left arm (fig 2).

CASE 3
This seven week old female infant died after attempted surgical correction of a common arterial trunk. There was the usual atrial arrangement with concordant atrioventricular connection and a subtruncal ventricular septal defect with an overriding quadricuspid truncal valve. The pulmonary arteries originally had separate orifices from the posterior
aspect of the common arterial trunk. The aortic arch was right sided and its branches were a mirror image of the normal arrangement. No arterial duct or ligament was present.

The left sided brachiocephalic vein descended to the left of the ascending aorta and then passed behind it to the right side, below the aortic arch, to open into the left medial aspect of the superior caval vein between the entry of the azygos vein and the junction with the right atrium (fig 3).

Fig 3 Photograph and diagram of case 3 showing the anomalous left brachiocephalic vein (ALBV) passing below the right aortic arch (RAA) before entering the superior caval vein (SCV). The photograph shows the vessels after surgical correction of the common arterial trunk (CAT), with the insertion of a pulmonary conduit (PC). The diagram shows the original state before operation. Ao, aorta; PA, pulmonary artery.

REVIEW OF PUBLISHED REPORTS
The true incidence of the anomaly is difficult to assess. The veins are rarely studied in detail at necropsy. Careless neck incisions, removal of the sternum, or opening of the pericardium may cause bleeding from the veins, which then collapse and become very difficult to see. Removal of the heart often damages or destroys the veins. Angiography does not usually call for visualisation of the right brachiocephalic vein. Increased routine use of suprasternal echocardiography, however, should readily reveal the anomaly if such a possibility is recognised.

A detailed angiographic study of the brachiocephalic veins of over 1500 people showed only one example of a subaortic vein,1 while another angiographic study of the superior caval veins in 510 patients with congenital heart disease showed no such anomalies.3 Sherman found two examples among 503 specimens of congenitally malformed hearts3 and we have encountered three cases out of approximately 2000 congenitally malformed hearts in the combined collections of the Brompton and Killingbeck Hospitals. These combined figures give an incidence of six cases out of about 2500 necropsy cases with congenital heart disease, or approximately one in 500. Smallhorn et al found seven cases echocardiographically but the total number of patients examined in the series is not stated.5

The incidence in individuals without cardiac malformation is presumably very much lower. In a personal necropsy series, LMG found no subaortic brachiocephalic veins in approximately 5000 individuals without malformed hearts.

In none of the reported cases has it been suggested that the venous anomaly had any adverse physiological or clinical effect. Therefore the age at death or diagnosis was relevant only to associated or unassociated conditions. The ages ranged from one day to 74 years (table). In those cases with congenital heart disease, the mean known age was approximately three years (excluding a 74 year old man with an anomalous connection of the right upper pulmonary vein). In cases with normally formed hearts, the mean age at death was 53 years, supporting the view that the abnormality is clinically benign.

The sex of 15 of the 25 cases was known. Males outnumbered females by 11 to 4. Those cases with congenital heart malformation showed no significant sex difference but all eight cases with normally formed hearts were male.

Features common to tetralogy of Fallot were present in most of the 14 cases with major cardiac malformations. Thus a ventricular septal defect was reported in eight cases and may have been present in another six reported by Smallhorn et al in which intracardiac malformations of unspecified types were found by echocardiography.5 A right sided aortic arch was noted in ten cases and tetralogy of Fallot, pulmonary stenosis, pulmonary atresia, or unspecified abnormalities of the right ventricular outflow tract were present in 11 of the 14. Other malformations were: one example of double outlet right ventricle, one common arterial trunk, and an anomalous isolated subclavian artery arising from an arterial duct (see table). There were minor vascular anomalies in three additional cases in which the heart itself was normal. A 74 year old man had anomalous drainage of the right superior pulmonary vein into the superior caval vein; a 19 year old man had a small subthyroid transverse venous anastomosis,10 and a man of 66 had minor abnormalities of the jugular vein.12 There were no instances of associated non-cardiovascular malformations.

Three distinct anatomical varieties of the malformation can be recognised according to the relation of the anomalous vein with the arterial duct or ligament (fig 4). The presence and position of this structure
Anomalous subaortic position of the brachiocephalic (innominate) vein

Anomalous subaortic position of the brachiocephalic (innominate) vein

An

...r

Artic...n

Ascendng

orta

Pumory

artery

Superior
cova

Left

brachiocephalic

- Azygs

Aor

/Pulmonary

artery

duct

Fig 4  (a) Diagram showing the positions of the normal (N) and the three variants of the anomalous left brachiocephalic vein (1, 2, 3) in relation to the 4th and 6th left aortic arches as seen from the left side. (b) Diagrams of the normal and abnormal variant venous pathways as seen from the front, with accompanying embryological diagrams.

Discussion

During normal development, the primordia of the systemic veins first appear as paired anterior and posterior cardinal veins that unite on each side to form a common cardinal vein (or cuvierian duct) that opens into the primitive sinus venosus. The anterior cardinal veins extend headwards to the junctions of the internal jugular veins and subclavian veins on each side, but, during subsequent development, most of the left anterior cardinal vein disappears. The venous drainage from the left side of the head and

was specified or illustrated in 13 cases. In ten, the vein passed anteriorly to the arterial duct or ligament to lie between the underside of the aortic arch and the pulmonary arteries. In two cases, the vein passed posteriorly to the arterial duct or ligament, while in the remaining case it passed below the left pulmonary artery. Three further cases have been described in which the left brachiocephalic vein passed behind one or more of the main aortic branches but was normally related to the aortic arch.

This condition is distinct from the anomalous venous pathway that is the subject of this review.
The subaortic vein enters the superior caval vein below the insertion of the azygos vein and it thus connects the left anterior cardinal venous component to the right common cardinal venous component. Normally, the left brachiocephalic vein enters the superior caval vein above the insertion of the azygos vein and the connection is between the left and the right anterior cardinal components. The anomalous vein, therefore, is embryologically distinct from the normal left brachiocephalic vein as well as having a different spatial pathway and different anatomical relations. In consequence, the question of terminology could be debated at length. But, because the ultimate connections at either end are identical, it seems reasonable to refer to the structure as an anomalous brachiocephalic vein.

Although not of clinical importance, its recognition during clinical investigation raises the possibility of an association with other malformations, especially right aortic arch, ventricular septal defects, and right ventricular outflow obstruction. Care must be taken not to confuse the structure with other arteries or veins—notably, the central pulmonary arteries in the presence of the pulmonary atresia or a venous confluence in anomalous pulmonary venous connection. From the surgical viewpoint, the anomalous brachiocephalic vein may hinder access to the arterial duct during ductal ligation, or obscure the field in the construction of a subclavian to pulmonary shunt. Dissection in the region of the duct must be performed carefully to avoid damaging a subaortic vein.

Fig 5  Diagrams showing embryological derivations of the normal and anomalous left brachiocephalic veins. (a) Early embryological patterns. (b) Normal definitive pattern showing the left brachiocephalic vein entering the right anterior cardinal vein component (above the azygos vein). (c) Abnormal definitive pattern showing the left brachiocephalic vein entering the right common cardinal vein component (below the azygos vein).
Anomalous subaortic position of the brachiocephalic (innominate) vein

We thank Dr Wilfred Nikolaizik for his valuable assistance in translating some of the references.

This study was supported, in part, by the British Heart Foundation.

References

Anomalous subaortic position of the brachiocephalic (innominate) vein: a review of published reports and report of three new cases.

L M Gerlis and S Y Ho

Br Heart J 1989 61: 540-545
doi: 10.1136/hrt.61.6.540

Updated information and services can be found at:
http://heart.bmj.com/content/61/6/540

These include:

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/