Congenital Communications of the Right Pulmonary Veins with the Azygos Vein

Report of a Case with Surgical Correction

By Howard N. Anderson, M.D., Warren G. Guntheroth, M.D., Loren C. Winterscheid, M.D., and K. Alvin Merendino, M.D.

PULMONARY venous drainage into the azygos vein is a rare type of anomalous pulmonary venous cardiac return. Brody found three cases reported in the medical literature to 1942 and added another case. More recently, this subject has been reviewed by Stecken and Beyer, who diagnosed three cases by tomogram and confirmed the diagnosis in one of these patients by angiocardiography.

To our knowledge, surgical correction of pulmonary venous drainage into the azygos vein has not been reported and for this reason it was thought that the following case report would be of interest.

Case Report

P., a 6-year-old, 34-pound white girl, was referred to the University Hospital for evaluation of a cardiac murmur first noted at age 2 years. She was essentially asymptomatic and had never been cyanotic. Physical examination revealed a small child with abnormalities confined to the heart. The point of maximal impulse was at the lower left sternal border. There was a grade-II/VI systolic murmur heard in the right first and second interspaces adjacent to the sternum, a grade-II/VI systolic murmur and a grade-I/VI diastolic murmur were heard along the left sternal border. An electrocardiogram revealed incomplete right bundle-branch block, right axis deviation and right ventricular hypertrophy. A cardiac series revealed a moderately enlarged right atrium and ventricle and increased pulmonary vascular-ity. There was a prominent, rounded bulge in the right upper mediastinum, thought to represent either enlarged lymph nodes or an anomalous pulmonary vein (fig. 1). (The aorta was located on the left on the basis of tracheal and esophageal position.) Cardiac catheterization revealed an oxygen step-up of five volumes per cent between the inferior vena cava and right atrium, a high saturation of the superior vena caval blood, and no right-to-left shunt. The pulmonary blood flow was twice the systemic blood flow. Right-sided pressures were essentially normal. Pulmonary vascular resistance was normal. A venous angiocardiogram revealed that all of the pulmonary venous drainage from the right lung entered the azygos vein, which in turn entered the superior vena cava (figs. 2 and 3).

Figure 1

Preoperative roentgenogram showing a prominent, rounded bulge in the right superior mediastinum, right-sided cardiac enlargement, and prominent pulmonary arteries.

From the Departments of Surgery and Pediatrics, University of Washington School of Medicine, Seattle, Washington.

Supported by Research Grant HE 03379, Postdoctoral Fellowship HPD-19,016, and Heart Training Grant 2 TTI HE 5194-06, U.S. Public Health Service.

Circulation, Volume XXX, September 1964

439
cardiogram. The operation was rapidly terminated after enlarging the atrial septal defect.

The patient's convalescence was uneventful. Because of concern that the hepatic vein emptied directly into the atrium and that the inferior vena caval return may have been contributing to the large azygos vein flow noted during open cardiotomy, a venogram was carried out. No anatomic evidence was noted to substantiate this thesis.

On June 20, 1962, the patient was re-explored and the same findings were observed. The only variation in technic was the placement of the superior venous catheter into the right innominate vein from the jugular via a small cervical incision. All systemic chest wall veins emptying into the azygos vein were doubly ligated and divided. A woven Dacron half-tunnel was utilized to channel all azygos blood into the left atrium via the atrial septal defect (fig. 4). This created some narrowing of the lumen of the superior vena cava. Consequently, this was widened by suturing a diamond-shaped piece of woven Teflon at a strategic location in the superior vena cava. The remainder of the operation was uneventful.

Re-examination 1 year postoperatively revealed the patient to be much more active physically than preoperatively. No cardiomegaly was present on physical examination. There was a grade-I/VI systolic murmur at the third left interspace. Chest films revealed little change in cardiac size. The prominent, rounded area of the right upper mediastinum, which represented the distended azygos vein, was no longer visible and pulmonary vascu-

**Figure 2**

*Early phase of venous angiocardio gram. A dilution defect is noted in the right superior portion of the superior vena cava at the entrance of the azygos vein.*

**Operation**

On October 27, 1961, the first open operation with extracorporeal support was performed via the right fourth intercostal space anteriorly. There was an enlarged right atrium, superior vena cava, and a huge azygos vein that emptied high in the superior vena cava close to the junction of the right and left innominate veins. No additional vessels were noted entering the superior vena cava.

The superior caval catheter was placed through the right atrium into the right innominate vein. The drainage via the left innominate vein was retrieved by retrograde drainage when the tourniquet on the cavae was occluded distal to the azygos vein. With the right atrium opened, a patent fossa ovalis was noted. About 300 to 500 ml. of blood per minute returned via the azygos vein. There was concern that other venous channels not easily visualized might be contributing to this flow. Additional dissection revealed none. At this point, however, there was a serious progressive deterioration of the patient's electro-

**Figure 3**

*Late phase of venous angiocardio gram showing right pulmonary veins draining into the azygos vein.*
larity was decreased from that noted preoperatively (fig. 5). Arterial saturation was completely normal, 98 per cent.

**Discussion**

Anomalous pulmonary venous drainage via the azygos vein is rare. With one exception all reported cases have occurred in the female. Of interest is the fact that it occurs in a variety of anatomic forms. There are cases of partial drainage from the right lung, total right pulmonary venous drainage, and total pulmonary venous drainage (i.e., both right and left). This lesion appears to be associated frequently with other congenital cardiovascular anomalies and abnormalities of lung genesis and development. It seems to have been diagnosed only twice prior to the authors’ case. Table 1, modified from Stecken and Beyer, describes the cases recorded in the medical literature to 1963 that have been proved by angiocardiography, operation, or autopsy.

The surgical correction of this lesion is not difficult. However, there can be special problems in venous cannulation for bypass due to the proximity of the azygos orifice to the confluence of the right and left innominate veins to form the superior vena cava. This may require jugular cannulation in the neck for satisfactory venous return from the upper body during bypass. In this patient, the intercostal veins were emptying into the azygos in their normal pattern. Obviously, all systemic venous channels emptying into the azygos must be divided and ligated. If this is not done, the azygos flow of oxygenated venous blood when rechanneled into the left atrium is contaminated by desaturated systemic venous blood. In short, while correcting the left-to-right shunt, one would at the same time be creating a right-to-left shunt. Therefore, a careful search with division of all systemic venous connections to the azygos is necessary. While one might guess that if this advice were ignored, the right-to-left shunt would be of little moment, it can only be stated from the experience of the first operation in this patient that venous return from the azygos was considerable. Unfortunately, the surgeon did not have sufficient presence of mind to occlude temporarily the right pulmonary artery so that the contribution to total azygos flow contributed by the systemic flow could be separately determined. Normally,
this volume is approximately 20 per cent of the total body flow.

At surgery, one will be sorely tempted to divide the intercostal veins, temporarily occlude the pulmonary artery, ligate and divide the azygos vein at its junction with the superior vena cava, and anastomose the proximal azygos to the left atrium. This was considered at the time of surgery. It was thought to be feasible but was discarded as a possible solution in this case. The decision not to do so was conditioned by the fear that due to the normally low intraluminal pressure in the vein, a small error in vein placement might result in kinking with congestive venous infarction of the right lung. Rather than risk this complication, a more familiar procedure was used.

**Summary**

A case of total pulmonary venous drainage of the right lung into the azygos vein, surgically corrected, has been reported.

There have been 10 previous cases of vary-

| Table 1 |
|-----------------|-----|-----|-----------------|------------------|------------------|
| **Pulmonary Venous Drainage into Azygos Vein** |   |   | Veins to azygos | Other anomalies | Confirmation |
| **I  Partial drainage of one lung** |   |   |   |   |   |
| Guillabert³ | 1859 | 55 yr. | M | Right middle lobe | Patent foramen ovale; right upper lobe veins to superior vena cava | Autopsy |
| Brody¹ | 1942 | 56 yr. | F | Right upper lobe | Agenesis of the left lung; one lobed right lung | Autopsy |
| Kjellberg⁸ | 1955 | 26 yr. | F | Two right upper lobe veins | Absent inferior vena cava; anomalous left innominate vein; pulmonary sequestration | Catheterization, angiocardioagram, and operation |
| Stecken² | 1963 | 18 yr. | F | One right lower lobe vein | Right upper lobe veins to inferior vena cava | Angiocardiogram |
| **II  Total drainage of either right or left lung** |   |   |   |   |   |
| Theremin⁴ | 1884 | 11 da. | F | Right lung | Agenesis of the left lung; one lobed right lung | Autopsy |
| Theremin⁴ | 1884 | 4 mo. | F | Right lung | Agenesis of the left lung; two lobed right lung | Autopsy |
| Shepherd⁶ | 1890 | 30 yr. | F | Right lung | Arterial anomalies | Autopsy |
| Hurwitz⁷ | 1937 | 7wk. | F | Left lung | Agenesis of the left lung; one lobed right lung | Autopsy |
| Authors' case | 1964 | 6 yr. | F | Right lung | Patent foramen ovale | Catheterization, angiocardioagram, and operation |
| **III  Total drainage of both right and left lung** |   |   |   |   |   |
| Muira⁵ | 1889 | 6 mo. | F | Right and left lungs | Patent ductus arteriosus; patent foramen ovale; superior vena cava entered left atrium; arterial anomalies | Autopsy |
| Edwards⁸ | 1953 | 10 yr. | F | Right and left lungs | Cor bilocular; subpulmonic stenosis | Autopsy |
ing degrees of partial right, total right, to total (bilateral) pulmonary venous drainage into the azygos vein. The authors’ case is the third to be diagnosed during life and apparently the first case to be successfully corrected. With one exception, all previously reported cases have occurred in females.

A review of previous cases and surgical considerations in the authors’ case have been presented.

References
3. Guillabert, V.: Montpellier méd. 3: 241, 1859. Quoted by Brody1 and by Stecken.2
4. Theremin, E.: Rev. mens. mal enf. 2: 554, 1884. Quoted by Stecken2 and by Hurwitz.7

Observations on Treatment
By Richard Bright—1827

In the foregoing statements it has been my great object to establish the fact, that certain dropsical affections depend more on the derangement of the kidneys themselves than has generally been supposed; and that the albuminous nature of the urine frequently points out the particular cases in which these organs are the seat of disease. I wish that I were now able to add any thing completely satisfactory to myself with regard to the mode of treating these diseases of the kidney. It will be very obvious from a review of the cases I have cited, that they sometimes present difficulties so formidable as to defy the ordinary means of cure; indeed I am inclined to doubt whether it be possible, after the decided organic change has taken a firm hold on the kidney, to effect a cure, or even to give such relief as may enable the patient to pursue for a few years the occupations of life; where, however, the mischief is less rooted, we may undoubtedly do much. In the treatment of the disease, as it occurs in sudden attacks of anasarca from intemperance and exposure, in its early stages, and before organic changes have taken place, we have two distinct indications to fulfil;—we have to restore the healthy action of the kidney, and we have to guard continually against those dangerous secondary consequences which may destroy the patient at any period of the disease.

The two great sources of casual danger will be found in inflammatory affections, more particularly of the serous, sometimes of the mucous membranes, and in the effusion of blood or serum into the brain, and the consequent occurrence of apoplexy.—Original Papers of Richard Bright on Renal Disease. Edited by A. Arnold Osman. London, Oxford University Press, 1937, pp. 71-72.
Congenital Communications of the Right Pulmonary Veins with the Azygos Vein: Report of a Case with Surgical Correction
HOWARD N. ANDERSON, WARREN G. GUNTEROTH, LOREN C. WINTERSCHEID and K. ALVIN MERENDINO

Circulation. 1964;30:439-443
doi: 10.1161/01.CIR.30.3.439
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1964 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/30/3/439

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/