LIKE ALL GAUL, the story of the intriguing anomaly total anomalous pulmonary venous drainage (TAPVD) is divided into three parts: (1) the 1950s, the age of discovery and definition; (2) the 1960s, the period of diagnosis; and (3) the 1970s, the era of definitive treatment. Because of its unusual features and its perplexing challenge to successful management, this malformation has enjoyed an attentive interest far out of proportion to its infrequent incidence.

Fortunately it is rare, for its effects are so devastating when it causes symptoms of respiratory distress, hypoxia, or of cardiac failure in babies. In three large series, each with 70-75 patients, mainly infants, that were reported on in 1968-70 but spanned 15-20 years of experience, mortality rates were 75-89% in the first year of life, with medical or surgical treatment.1-3 Perhaps 10% of patients with this anomaly are brought to medical attention after the age of 1 year, and they have a much more benign situation, resembling a large atrial septal defect in its manifestations, and more successful surgical results.

In the 1950s the range of severity, embryology, and classification were defined. The most generally accepted classification is that of Darling,4 who divided the anomalies of drainage into four groups according to their point of juncture with the right atrium or a tributary thereof. In decreasing order of frequency, these are supracardiac, cardiac, infracardiac, and mixed.

In the 1960s precise diagnosis of the lesion became a reality when physiologic assessment by cardiac catheterization was combined with selective angiocardiography for demonstration of the anomalous course and point of entry of the pulmonary veins. It became evident that there were two kinds of sick infants with TAPVD: (1) those who presented around the age of 6-8 weeks with mild cyanosis, great enlargement and hyperactivity of the right side of the heart, and large left-to-right shunts; and (2) those who presented in the first days of life with tachypnea and cyanosis, small hearts, and near-average pulmonary blood flow, but on chest roentgenogram a stippled vascular pattern throughout the lung fields. Prognosis for the first group, who had no elevation of pulmonary venous or pulmonary capillary pressure, and whose pressure in the pulmonary artery was less than 50% of the systemic pressure, was better than for the second group, which was characterized by marked pulmonary arterial hypertension.

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often to greater than the systemic level, due to obstruction to pulmonary venous return at some point in the anomalous course into or out of the left heart.

Obstructed pulmonary venous return was first recognized in patients with infradiaphragmatic drainage into the portal vein or ductus venosus, but then it became evident that patients with supracardiac connection could also be obstructed.5-7 One mechanism of obstruction is constriction of the anomalous vertical vein as it passes posterior to the left pulmonary artery and anterior to the left main bronchus. Second, since in this malformation of TAPVD the point of access to the left atrium and systemic circulation is the patent foramen ovale or a defect in the atrial septum, too small an opening at this level could obstruct pulmonary venous return. Rashkind and Miller postulated that balloon septostomy in newborns with obstruction at the interatrial communication might relieve this situation.8

Third, because the left atrium and ventricle appear so much smaller in this malformation than their counterparts in the right heart, there has been speculation that hypoplasia of these structures may be responsible for pulmonary venous congestion and edema preoperatively and may account for the deaths after repair.1 The recent left heart volume studies of Graham et al.9 suggest that this is not so, however, and current surgical successes would also deny that the left side of the heart is critically small.10-12

As recently as 1968, Mustard et al. wrote regarding this lesion that “the risk of surgery is considerable and indeed, survivors in the first six months of life when surgery is so urgently needed still remain extremely rare.”1 The beginning of the 1970s has already seen this change, so that the saga of a rare, ever challenging, and often frustrating anomaly can end as a success story. The article in this same issue by the group from Great Ormond Street tells of the learning processes in recognition and both medical and surgical management that led to their improved results over the past 7 years in the surgery on 37 sick infants. During this period there were 13 survivors; the best survival rate was in the nine babies of 11 who had supracardiac drainage.10 Gersony et al. reported in 1970 that of 10 consecutive babies operated upon, seven with supracardiac or cardiac drainage survived and were doing well on follow-up.11 Barratt-Boyces and associates that same year reported that six babies with TAPVD survived operation, performed in two of them at 8 and 9 days, respectively.12

Many interrelated factors have contributed to this success story. Among these are not only consummate surgical skill but also the improved understanding of the pathophysiology of the anomaly and of the patient at high risk. Important too are the many pediatric, cardiologic, surgical, anesthesiologic, and nursing advances that have developed since attention was focused on the newborn with heart disease by cardiac centers fully staffed and equipped to provide expert care 24 hours a day.13 The system requires expertise at every point: (1) early recognition of the dyspneic or cyanotic infant, (2) prompt transport with provision of warmth and oxygen to a cardiac center, (3) immediate evaluation of the condition and correction of problems such as respiratory or metabolic acidemia and cardiac failure, (4) performance of cardiac catheterization with selective injection of contrast medium in appropriate amounts for definition of malformation, (5) early operation with utilization of the technics of anesthesia and circulatory support appropriate to that center, (6) intensive postoperative management by the team with special attention to monitoring and cardiorespiratory therapy, and (7) continued long-term cardiologic supervision to assess the results of surgery and to promote good health and activity.14

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Total Anomalous Pulmonary Venous Drainage Success Story at Last
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