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Anomalies of the Systemic Venous System*

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Minor variations in venous return are very common. Major anomalies, however, are less frequent and mainly confined to the inferior and superior vena cava and to the pulmonary veins.

Recent developments in the diagnosis and treatment of cardiovascular disorders have brought anomalies of the systemic veins to the attention of the cardiologists and the thoracic surgeons.

The classification of the anomalies of the systemic venous system is based on the embryologic principles. This classification includes, anomalies of the cardinal venous system, anomalies of the inferior vena cava and anomalies of the valves of the sinus venous (Table I). Total anomalous systemic venous connection is also to be considered.[1,2]

TABLE I
ANOMALIES OF THE SYSTEMIC VENOUS SYSTEM

I) Anomalies of the Cardinal Venous System
- Persistent left superior vena cava (LSVC) connecting to right atrium (normal formation of coronary sinus).
- Persistent left superior vena cava connecting to left atrium (failure of coronary sinus development).
- Right superior vena cava connecting to left atrium.
- Anomalies of the coronary sinus

II) Anomalies of the Inferior Vena Cava
- Infrahepatic interruption of IVC with azygous continuation.
- IVC connecting to left atrium.

III) Persistent Valves of the Sinus Venous

IV) Total Anomalous Systemic Venous Connection

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The present paper is a retrospective study intended to establish the incidence of the anomalies of the systemic venous return and to review the clinical and laboratory findings related to these anomalies.

Materials and Methods

87 subjects with anomalies of systemic venous return were found among 2000 patients hemodynamically studied in the last five years. Right ventricular angiography was used for the purpose of detection. The incidence was found to be 4 %. The patients ages ranged between 3 and 20 years. The mean age was 11 1/2.

Results

56 subjects had persistence of the left superior vena cava (LSVC) with the congenital cardiac defect (Table II). Four subjects had the left superior vena cava connected to the left atrium (7 %) (Figure 1). The LSVC drained into the right atrium in the remaining 52 subjects (93 %). Tetralogy of Fallot was found in 18 cases. This was the most common congenital cardiac defect in this group (Table III). Atrial septal defects and ventricular septal defects formed the second most important anomalies. There were 9 cases of each of the above septal defects. Other anomalies were rare. Only one case of each of the following congenital cardiac defects were found; Taussig-Bing malformation, endocardial cushion defect and pseudotruncus. In four cases LSVC was connected to the left atrium. The associated cardiac anomalies in these four cases are summarized in Table IV.

TABLE II

THE INCIDENCE AND THE DISTRIBUTION OF THE 87 SUBJECTS SHOWING ANOMALIES OF SYSTEMIC VENOUS RETURN FOUND IN HEMODINAMICALLY STUDIES 2000 CASES

The anomalies	Number	Incidence
Anomalies of the systemic venous return	87 cases	4 %
Persistent left superior vena cava	56	2,8 %
a) LSVC connecting to right atrium	52	(93 %)
b) LSVC connecting to left atrium	4	(7 %)
Anomalies of the inferior vena cava	30	1,5 %
Anomalies of the coronary sinus (Communication of coronary sinus with both atria)	1	0,5 %

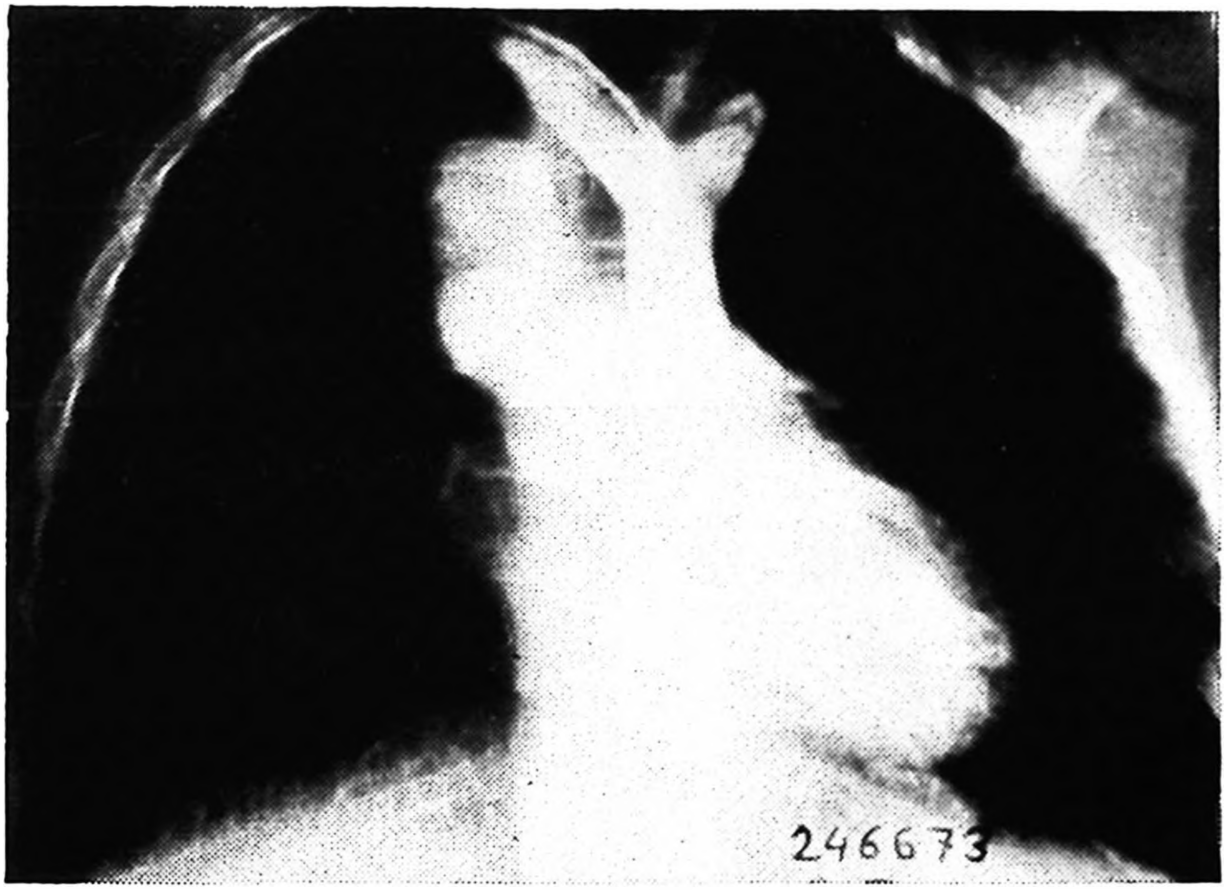

Figure 1
Left superior vena cava connected to the left atrium

TABLE III

ASSOCIATED CARDIAC ANOMALIES WITH LSVC

Congenital cardiac defect	Number
Tetralogy of Fallot	18
Atrial septal defect	9
Ventricular septal defect + pulmonary hipertention	9
Transposition, great arteries	2
Dextrocardia	2
VSD + ASD	4
Tricuspid Atresia	5
Taussig-Bing malformation	1
Endocardial cushion defects	1
Pseudo truncus	1
Isolated	1

TABLE IV

ASSOCIATED CARDIAC ANOMALIES WITH LSVC CONNECTING TO LEFT ATRIUM

Congenital Cardiac Defect	Number
Tetralogy of Fallot	1
ASD + VSD	1
VSD + PS	1
Single Ventricle	1

Four subjects were diagnosed as having absence of the superior vena cava (Figure 2). In twenty eight cases absence of the IVC and continuation of the azygos vein could be detected (Table V). The incidence was 1.4 % (Figure 3). One subject had the left vena cava without situs inversus (Figure 4).

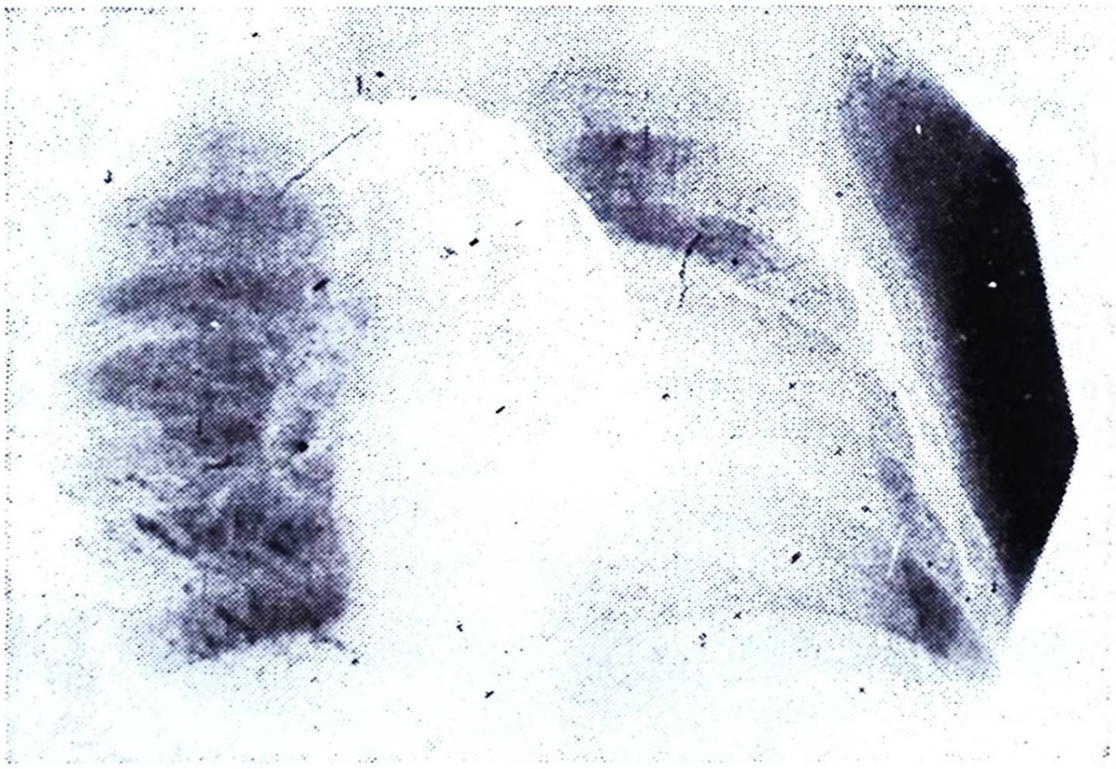

Figure 2

Absence of the superior vena cava

TABLE V

ANOMALIES OF THE INFERIOR VENA CAVA

Infrahepatic Interruption of IVC With Azygos Continuation	28
Left Vena Cava Instead of the Right Without Situs Inversus	1
Supracardinal and Subcardinal Veins	1

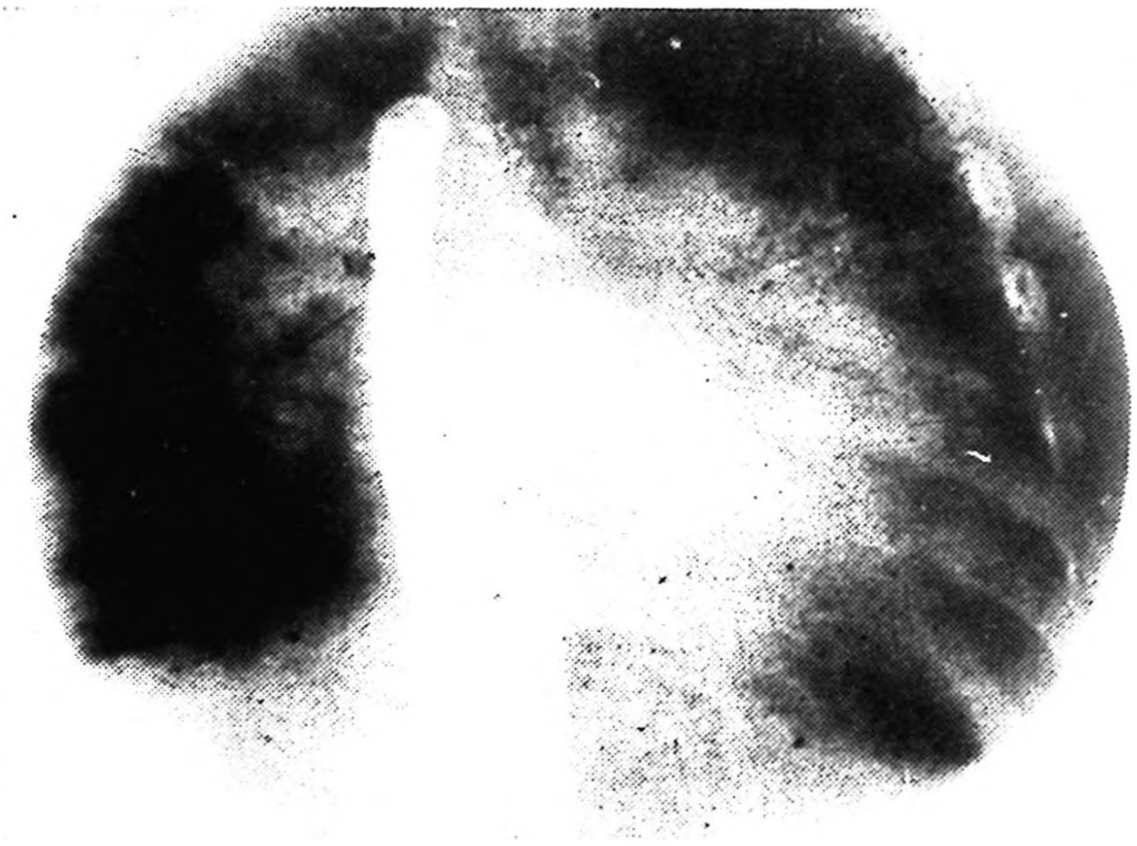

Figure 3
Absence of the IVC and continuation of the azygous vein

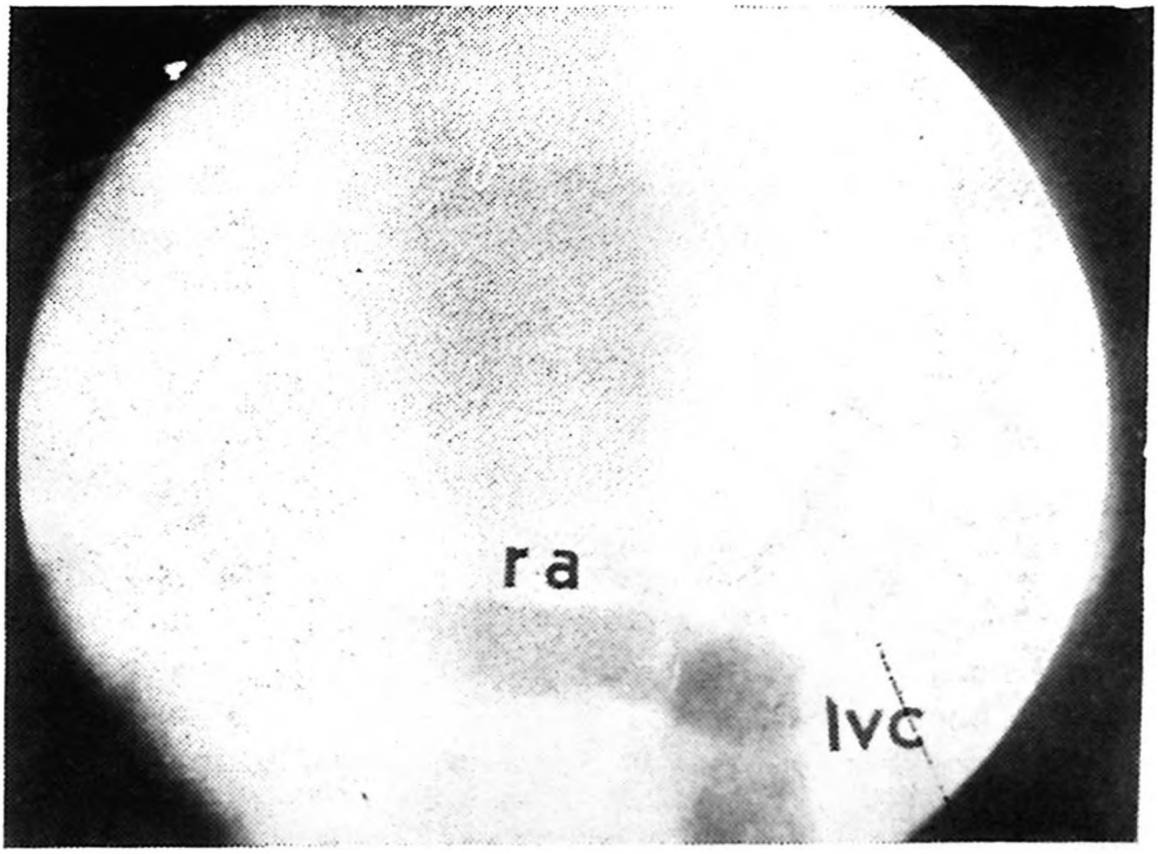

Figure 4
Left vena cava inferior

Only one patient had sub and supra cardinal veins (Figure 5).

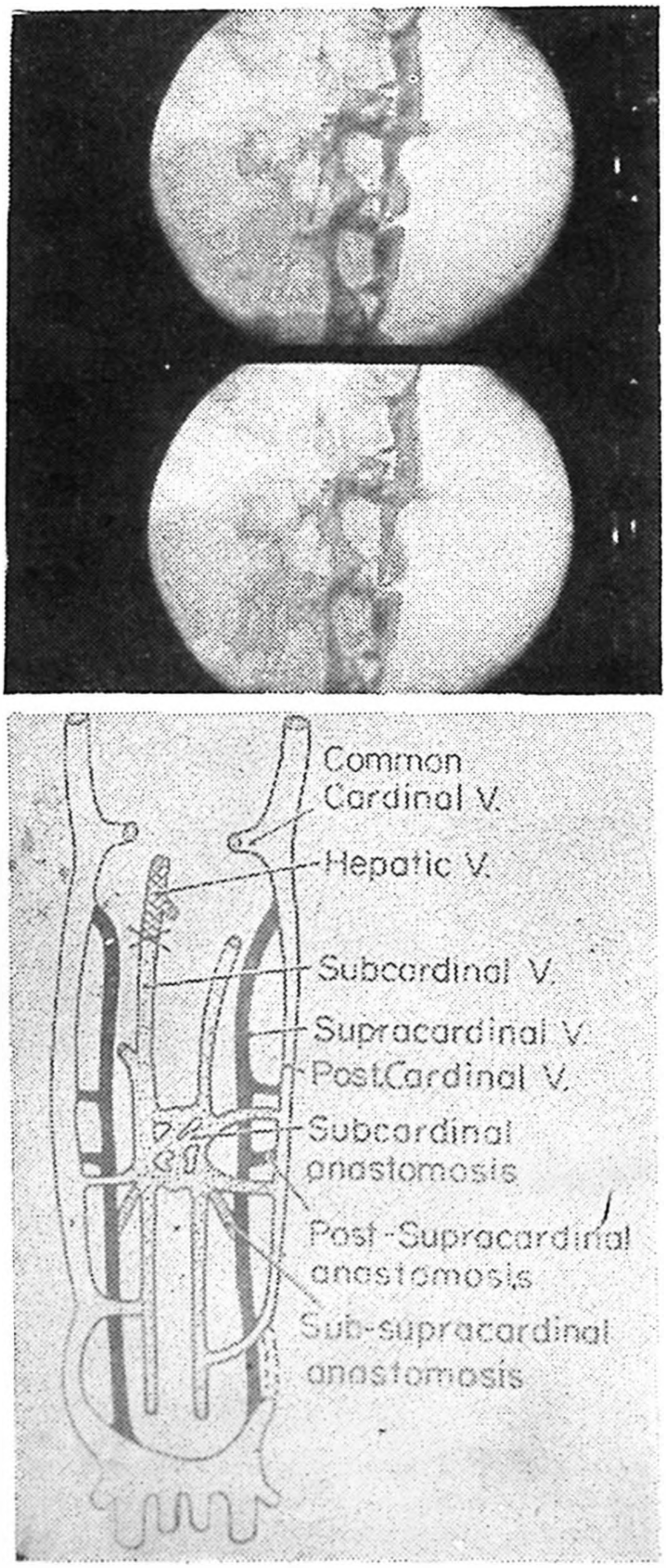

Figure 5

a-b) Sub and supra cardinal veins

Associated anomalies with the absence of the IVC are listed in Table VI. Six cases of tetralogy of Fallot were identified among the thirty patients with absence of IVC in the same group. There were four cases of situs inversus, two dextrocardia cases (with ASD + VSD + PS) and three cases with levocardia + VSD + ASD + asplenia and one with transposition of the great arteries.

TABLE VI

ASSOCIATED CARDIAC ANOMALIES WITH ANOMALIES OF THE INFERIOR VENA CAVA

Congenital Cardiac Defect	Number
Tetralogy of Fallot	6
ASD	3
ASD + VSD + PH	2
VSD + PH	1
AS + PDA	1
PS	1
AK + PDA	1
ASD + Single Ventricle	1
Endocardial Cushion Defect	1
Transposition of the Great Vessels + PS	1
Tricuspid Atresia	1
Situs Inversus Abdominalis + Levocardia	4
Situs Inversus Abdominalis + Dextrocardia	2
Situs Inversus Abdominalis + ASD + VSD + PS + Asplenia	2
Dextrocardia + ASD + VSD + PS	1
Dextrocardia + Single V. + PDA	1
Supracardinal and Subcardinal Veins	1

Discussion

The classification of the anomalies of the systemic venous system are based on the embryologic principles. These developmental aberrations may come up from 3 main systems as shown in Table I. Anomalies of the cardinal venous system included developmental aberrations of the right and left superior vena cava and the coronary sinus.

Failure of obliteration of the left anterior cardinal vein results in persistence of the LSVC. When the invagination fails to occur, coronary sinus does not develop, and LSVC connects directly to the left atrium.

This occurs rarely as an isolated defect occasionally in association with the defects of the atrial septum and commonly with primitive cardiovascular defects.[2,3]

Many variations in the formation of the IVC have been described. These include double IVC, left IVC. Anomalies of inferior vena cava occasionally cause problems for the abdominal surgeon. Infrahepatic interruption of the IVC with azygous continuation and IVC connecting to the left atrium are the important anomalies of the inferior vena cava.

In our series the only common anomaly of the superior cava system was persistent left superior vena cava. The incidence was found to be 2.8 %. 4000 unselected autopsies the incidence reported in the literature was 0.3 %.[4] In patients with additional cardiac defects the incidence of LSVC was higher and ranged from 2.8 % to 4.3 %.[5,7] Wood found persistent LSVC in 20 % of the cases with tetralogy of Fallot.[9] Tetralogy of Fallot was the most common defect among our cases, too.

The incidence of LSVC is high in patients with sinus venosus atrial septal defects. ASD and VSD also formed the second most important anomalies in our cases.

In 28 subjects we found absence of the IVC and continuation of the azygous vein. Anderson et al.[9] reported 15 patients with interruption of IVC with azygous continuation among 2500 cases of congenital heart disease studied by cardiac catheterization. Dupicis[10] et al. found higher incidence, 2.9 %, we found the incidence to be 1.4 %.

We concluded that since congenital systemic venous anomalies are frequently associated with congenital heart disease, a special attention should be paid to detect these venous anomalies, during catheterization, however, this data could be useful during the surgical intervention if needed.

Summary

Eighty-seven subjects with anomalies of systemic venous return were studied in detail. Angiography was used for the detection and the evaluation.

The patients were between 3 and 20 years of age. Twenty nine subjects had persistence of the left superior vena cava with congenital cardiac defects. Four subjects had left superior vena cava connected to the left atrium. Left superior vena cava drained into the right atrium in the remaining twenty-six subjects. The inferior vena cava was absent in

twenty-four subjects. One subject had left vena cava inferior instead of the right vena cava without situs inversus. Four subjects were diagnosed as having absence of the superior vena cava. Only one patient had sub and supra cardial veins.

It was emphasized that special attention should be given for the detection of these anomalies because they may be present an important problem during cardiac surgery and in the differential diagnosis of mediastinal masses.

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