CHIRURGIE CARDIAQUE / CARDIAC SURGERY

MULTIPLE VALVE SURGERY IN AN ADULT PATIENT WITH SITUS INVERSUS, DEXTROCARDIA AND RHEUMATIC HEART DISEASE

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ABSTRACT

Dextrocardia with situs inversus is a rare congenital disease. In the adult hood, its diagonsis via a rheumaticpolyvalvular disease is not frequent and its sutgical approach is not well codified. We report a case of a vietnamese of 47 years old to whom o mitral valve replacement (MVR) and an aortic and tricuspid vlavze repair were successfully performed.

Keywords : Destrocardia, Situs inversus, Rheumatic heart disease (RHD)

RESUME

La destrocardie à un situs inversus est une cardiopathie congénitale rare. A l'âge adulte, sa révélation par une polyvalvulopathie rhumatismale est particulière et sa prise en charge mal codifiée. Nous rapportons un cas d'ubne vitenamienne de 47 ans, chez qui un remplacement valvulaire mitral, une valvulopathie aortique et une annuloplastie ont été réalisés avec succès.

Mots-clés : *Destrocardie, Situs inversus, Cardiopathies rhumatismales*

Introduction

Rheumatic heart disease in children and adults remains a significant problem in Vietnam. The present case describes an unusual operation for acquired rheumatic heart disease in a patient with congenital situs inversus with dextrocardia, or mirror image dextrocardia. This is an underlying congenital visceroatrial situs condition with no anatomical, functional, or clinical dysfunction relating to the congenital condition. Familiarity with the topographical features of the heart position warranted an alternative surgical approach, which will be highlighted and described.

Clinical Case

A 47 year old Vietnamese woman was referred to Viet Duc Hospital in Hanoi, Vietnam for further evaluation and treatment of congestive heart failure secondary to Rheumatic Heart Disease (RHD). The patient initially presented to Uong Bi Hospital (in Quang Ninh province – Vietnam) and National Heart Institute – Bach Mai Hospital in Hanoi with a one month history of

progressive fatigue, dyspnea at rest, and intermittent chest pain. Medications included digoxin, diuretic, potassium supplement, and no anticoagulation. Past history was significant for bilateral ankle swelling at 12 years of age from suspected rheumatic fever. Patient was subsequently treated intermittingly for 30 years for presumed RHD. Clinical examination at Viet Duc hospital revealed exertional dyspnea, blood pressure 100/60 mm Hg, irregular pulse at 100 beats/minute, a right apical cardiac pulsation, a 3/6 systolic murmur with a diastolic rumble along the right lower sternal border, and radiation to the right axilla. There was bilateral pedal edema, bilateral cervical neck vein distension, and palpation/percussion of the liver edge 2 cm below the left costal margin.

The diagnostic evaluation included a chest roentgenogram (figure 1). This confirmed situs inversus with a left sided liver, dextrocardia, right aortic arch, and the stomach bubble on the right side. The cardiothoracic ratio (CT ratio) was 80%, the left atrium enlarged, and the lung fields congested.

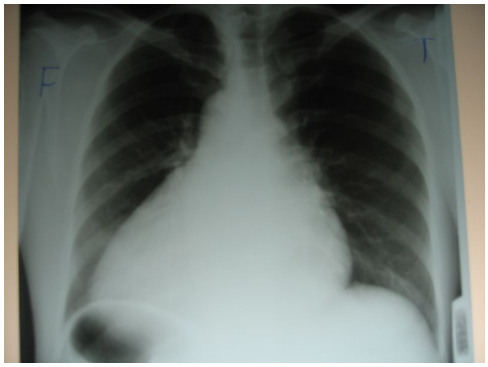


Figure 1. PA upright CXR with cardiac apex to the right, gastric bubble on right, liver on left side, and a right aortic arch.

The ECG showed atrial fibrillation with a ventricular rate of 90 beats/minute (**figure 2**). A 2D echocardiogram (2 D ECHO) revealed left atrial enlargement (50 mm); Left ventricular end diastolic dimension (LVEDD) of 65mm.; dilated right ventricle; ejection fraction (EF) of 50%; and an increased pulmonary artery pressure (40mmHg).

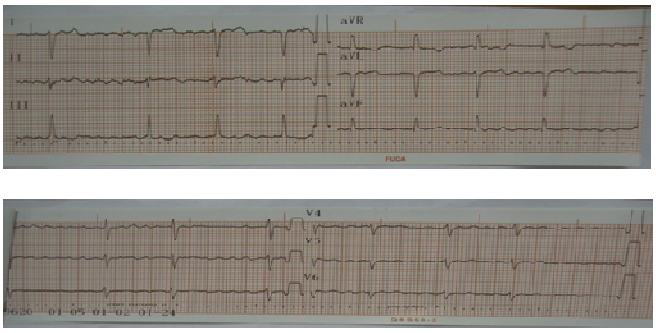
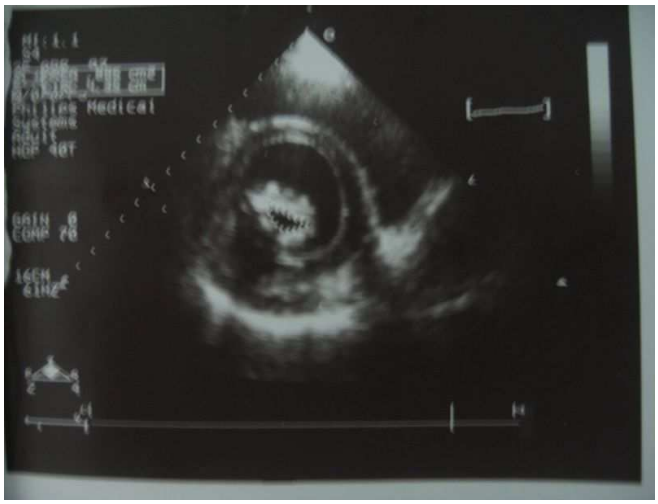


Figure 2. Atrial fibrillation; Right axis with negative forces in I, AVL, and V4,5,6; and biventricular hypertrophy.

There was calcific mitral valve stenosis with a mitral valve area (MVA) of 1.2cm 2, 3 plus mitral valve insufficiency, calcific aortic valve stenosis (aortic valve gradient (AVG) of 30 mm Hg), 2 plus aortic valve regurgitation, and moderate tricuspid insufficiency (**figure 3 a, b,c,d)**.



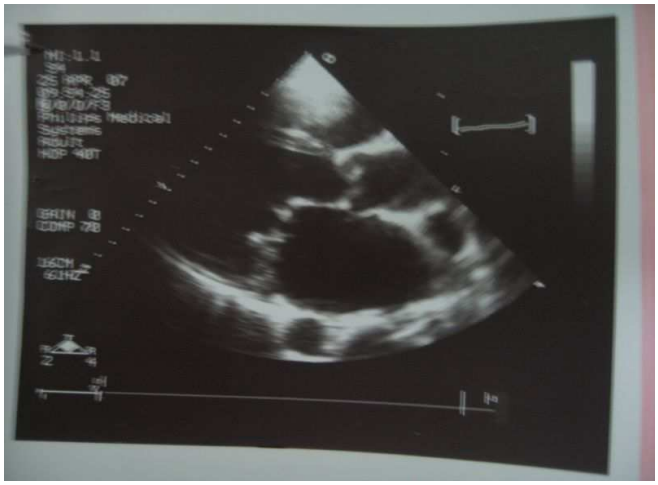


Figure 3(a)(b) Short and long axis views. See text.

The clinical features and diagnostic tests were consistent with significant rheumatic mixed and multiple valve disease (NYHA Class III), and associated situs inversus with dextrocardia. Corrective open heart surgery was recommended.

At operation a median sternotomy was performed. The topography of the heart revealed dextrocardia, with the anatomical right atrium and both vena cava on the left side **(figure 4a,b).**

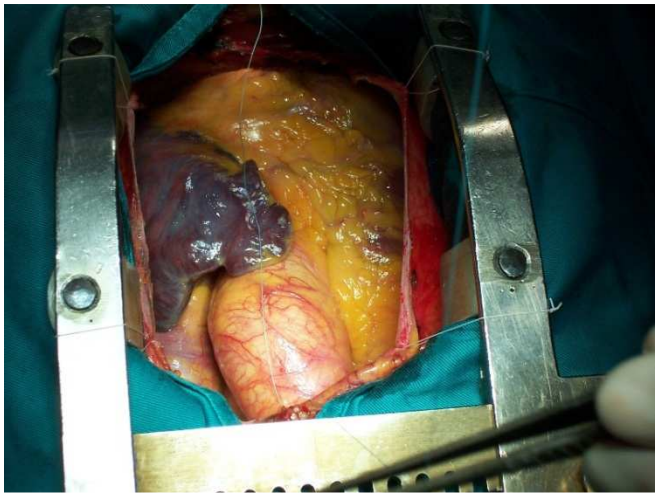


Figure 4 (a) Superior or cervical view from head of table. Anatomical right atrial appendage, right atrium, right ventricle, and both superior and inferior vena cava are on the left side of the patient

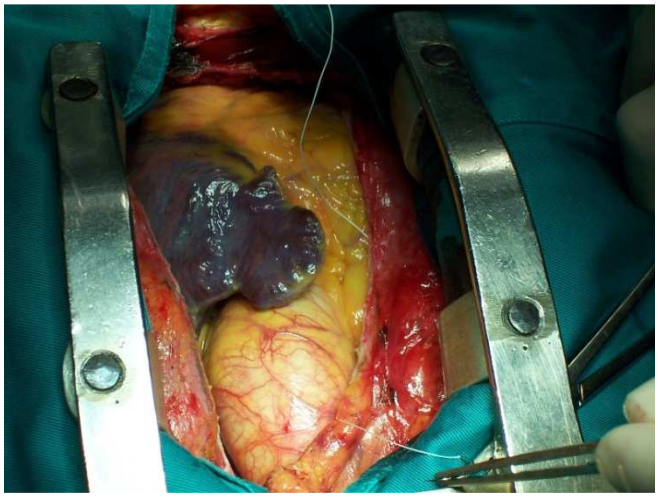


Figure 4 (b) Closer view of dilated ascending aorta, and dilated anatomical right atrium and ventricle on the left side

Bicaval venous and distal ascending aortic cannulation was performed from the patient's left side (**figure 5**).

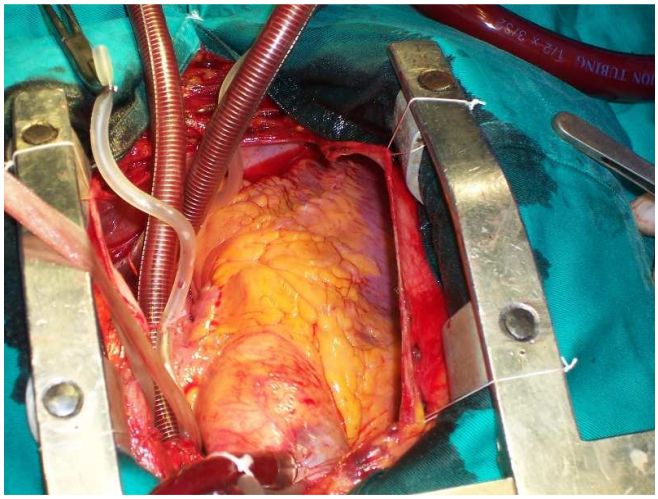


Figure 5 Bilateral caval cannulation from the left side, and distal aortic cannulation of right aortic arch.

Employing hypothermic (28 degree centigrade) systemic cardiopulmonary bypass, aortic cross-clamping, and cold blood cardioplegic arrest.
A right atrial and transeptal approach to the mitral valve was performed,revealing a stenotic mitral valve with calcification and retraction of both anterior and posterior leaflets causing both regurgitation and stenosis.A mitral valve replacement with posterior leaflet preservation was performed with a mechanical valve (St-Jude # 31). Next, a transverse ascending aortotomy approach to the aortic valve was performed from the left side (**figure 6**).

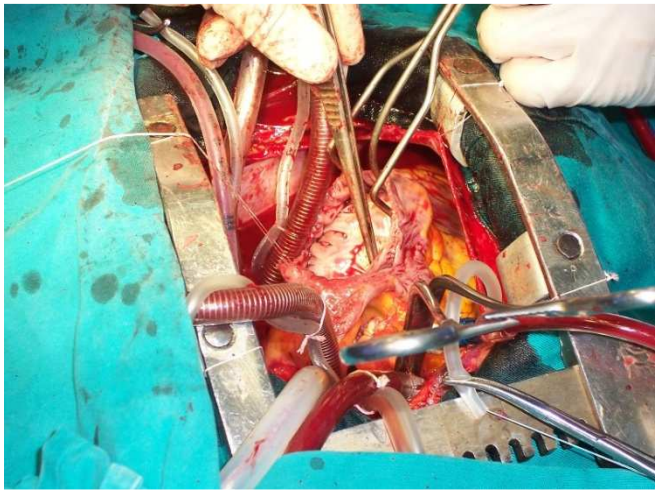


Figure 6. Right transatrial and septal approach to both tricuspid and mitral valves from the left side

The aortic valve was stenotic with calcified leaflets, consistent with rheumatic disease, characteristic in Vietnam). The aortic valve was replaced with a mechanical valve (St Jude # 21). The tricuspid valve was repaired with the De Vega procedure. There were no technical difficulties exposing the tricuspid, mitral, or aortic valves. At end of procedure, the heart reverted to sinus rhythm after the second electrical shock .
The postoperative course was uneventful, without the need for inotropic drug support. The patient was extubated on the fourth postoperative day. The patient remained in normal sinus rhythm. At one year follow-up the patient was NYHA Class I-II with decreased heart size (CT ratio) on CXR (**figure 7**).

Figure 7. Improved patient at one year clinical and diagnostic follow-up.

The follow-up 2D ECHO was improved: Left atrial 44 mm; left ventricular end diastolic dimension (LVEDD) of 52mm; right ventricle 30mm; ejection fraction (EF) of 58%; and pulmonary artery pressure 33mmHg.

Discussion

The prevalence of adolescent and adult CHD (ACHD) continues to increase worldwide in both the developed and developing countries (1). A growing number of this population will require surgical treatment (2). This group includes patients following previous palliative or corrective surgery, sequelae from previous surgery, patients with recognized or unrecognized ACHD without previous surgery, and patients with CHD requiring acquired heart disease operations. Guidelines have been published for the management of ACHD (3). Further guidelines have been published for the establishment of regional ACHD centers (4). The population range per center is recommended from 1 per 3 million to 1 per 10 million for the USA and Canada (4). No similar data or guidelines exist for emerging economies or developing countries.
Common clinical conditions in the ACHD group include cyanosis with secondary polycythemia, aortopulmonary collaterals, and varying degrees of myocardial dysfunction secondary to volume or pressure overload conditions (5).

Vida et al. (2) reviewed 2,012 adult CHD (18 years of age) requiring surgical treatment in the European experience. There were 4 groups: (1) Those surviving to adulthood, without previous cardiac surgery, and no irreversible heart or lung damage (75%); (2) Candidates for corrective surgery following previous palliative operation; (3) Patients with late complications or residual defects following previous surgery (23.1%); and (4) Patients requiring additional palliative surgery or heart transplantation (1.9%). Overall hospital mortality was 2%. Cyanosis, arrhythmias, and NYHA class III-IV were risk factors for mortality. Overall survival probability was 97% at 60 months, with 98.2% in the corrective group, 94.1% in the reoperation group, and 86.1% in the palliative group.
Few case reports are reported for ACHD, and associated acquired cardiac disease. ACHD and acquired coronary artery disease (CAD) is the most common condition encountered(6-8). Associated acquired valve disease with ACHD is rarely reported (9). The most common combined operation is coronary artery bypass graft (CABG), and repair of a congenital atrial septal defect (ASD) (10). The ASD or Patent Foramen Ovale (PFO), when recognized preoperatively, is usually repaired at the time of CABG. There are other acquired diseases anatomically associated with ACHD. Examples include endocarditis of congenital defects (eg.VSD, PDA, Coarctation),and Lutembacher's syndrome. The latter is a syndrome of rheumatic mitral stenosis in association with a congenital ASD (11). Neither example will be discussed, wheras RHD, because it is more prevalent in emerging economies or developing countries like Vietnam, deserves further discussion.

Of the 57,029 global mortality in 2002, cardiovascular disease (CVD) was the leading cause with 16,733 deaths (12-17). This included 7,208,000 deaths from ischemic heart disease, and 327,000 deaths from rheumatic heart disease (RHD). In the 15-29 year old age group there were 48,062 deaths in 2000 from RHD. RHD has declined significantly in the developed countries, but remains a significant problem in the developing countries or emerging economies. The global prevalence of RHD is estimated at 15-19 million. The incidence of mixed and multiple valve disease is more common in RHD than other acquired disorders, including degenerative valve disease. Specific indications for surgery are more difficult to establish(18).

The results of triple valve surgery for RHD continues to improve, especially in emerging economies or developing countries. Han et al. (19) from China, reported 871 patients from 1985-2005 with RHD undergoing triple valve surgery. The mean age was 42. The 30 day hospital mortality was 8%. Risk factors for mortality included ascites, NYHA Class IV, and decreased LV function. Long term cardiac survival was 75% at 5 years, and 63% at 10 years.Long term follow up remains difficult, as well as anticoagulation regulation.

Cardiac malposition is the most common generic term used to describe the
position of the cardiac apex in the right or left chest. The incidence of malpositions is 0.1-0.2/1,000 population with equal male/female distribution. Historically, a number of confusing classifications and terms have been used (20-26). **Figures (8,9,10,11)** summarizes a contemporary classification scheme. Situs inversus with dextrocardia is more common in the adult population given the <5% incidence of associated congenital heart defects (Table 1) (23,24).

Table1. Incidence of Extra and Intracardiac Anomalies associated with Dextrocardia (Modified from (23,24)

Situs Inversus with dextrocardia	
Normal heart	95%
Congenital lesion	5%
AV discordance	25%
Corrected TGA	20%
Complete TGA	30%
DORV	30%
PS/PA	50%
VSD	60%
Right aortic arch	80%
Situs Solitus with dextrocardia	
Normal heart	5%
Congenital lesion	95%
AV discordance	50%
Corrected TGA	50%
Complete TGA	10%
DORV	10%
PS/PA	60%
VSD	60%
Right aortic arch	5%

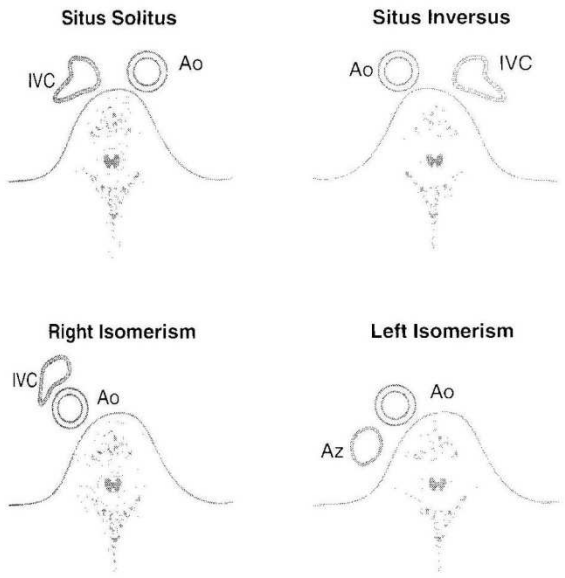


FIGURE 8 • Situs by echocardiography. Situs solitus is normal, and the aorta *(Ao)* and inferior vena cava *(IVC)* are symmetrically positioned adjacent to the spine. In situs inversus, there is a mirror image relationship. In right atrial isomerism, the IVC and Ao run together on either side of the spine. In left isomerism, there is azygos continuation of the IVC(Az) located retroperitoneally with the Ao.

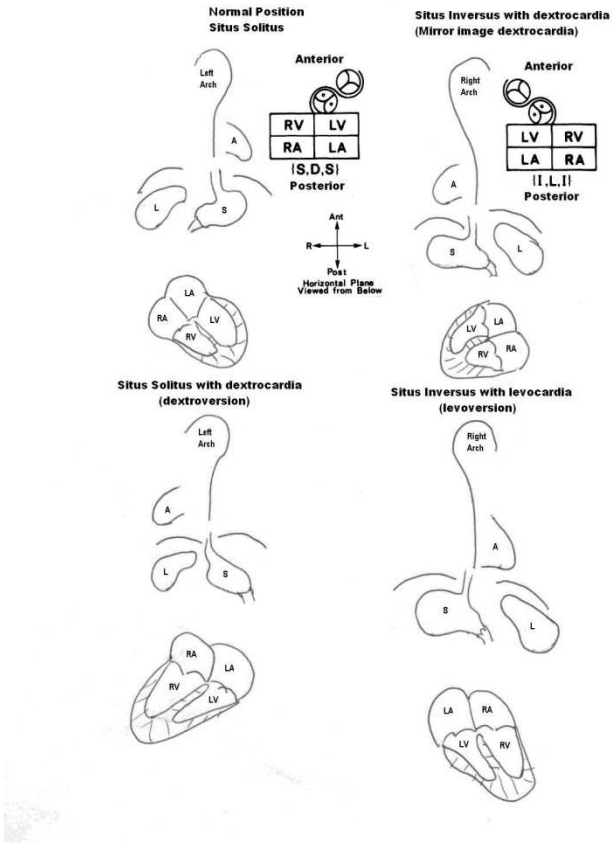


Figure 9 Composite classification of the four basic types of situs. Heterotaxy is not elaborated. (RA-right atrium; RV-right ventricle; LA-left atrium; LV-left ventricle; S-stomach; L-liver; A-apex) (Modified from Elliott et al. Invest. Radiol 1966;1:17.)

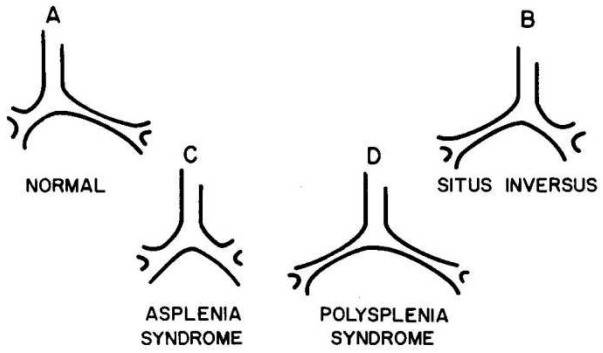


Fig. 10 Bronchial tree architecture as a guide to atrial situs. Diagramatic representation of the bronchi in situs solitus or normal *(A)* and situs inversus *(B)*. The morphologic right bronchus is wide and short and descends steeply, and the morphologic left bronchus is long, narrow, and descends more horizontally *(A)*. In situs inversus there is right-to-left reversal *(B)*. Bilateral morphologic right bronchi are seen with asplenia syndrome *(C)* and bilateral morphologic left bronchi are seen with polysplenia syndrome *(D)*. (From Rao PS, and Leonard T: Cardiology Digest 11:14, 1976.)

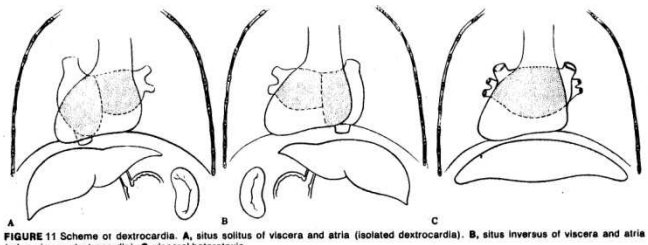


FIGURE 11 Scheme of dextrocardia. **A**, situs solitus of viscera and atria (isolated dextrocardia). **B**, situs inversus of viscera and atria (mirror-image dextrocardia). **C**, visceral heterotaxia.

Kartagner's syndrome is a rare condition in the adult that is characterized by situs inversus, chronic sinusitis, and bronchiectasis (27). This is a disease caused by a deficiency or inability of the pulmonary mucociliary clearance mechanism.

Acquired conditions of malposition are unusual. Previous surgical pneumonectomy, bilobectomy, or thoracoplasty can cause mediastinal shifts with movement of the heart to the midline or opposite chest. In the adult, all acquired causes of dextrocardia must be ruled out.

Heterotaxy syndromes or visceral heterotaxy are rare in adolescense or adults, since they are have a higher incidence of complex defects and eary lethality. Over 79% of heterotaxy patients die within the first year. A contemporary definition has been proposed by Stella Van Praagh (28).

"Visceral heterotaxy (from the Greek word heteros, meaning other, and taxis, meaning order) is a syndrome characterized by inconsistency of the situs of the situs of the thoracic and abdominal viscera and frequently by the preservation of the early embryonic symmetry of the liver and some of the systemic veins."

Further, this syndrome has a high incidence of congenital heart defects, and bilateral isomerism, i.e. right or left. Right sided isomerism is characterized by

a right bronchial and lobar pattern, and associated asplenia, whereas left sided isomerism includes a left bronchial and lobar pattern with polysplenia ( ).

This syndrome has been further reviewed by Winberg(26). He classifies 3 forms of visceroatrial situs. Situs solitus is the normal or most common. Situs inversus with dextrocardia is the mirror image of situs solitus, as is the present case. Combinations of situs solitus and inversus are termed situs ambiguous or heterotaxy syndrome, since elements of both may be present, e.g. midline liver (figure 11 c).

Contemporary results with complex operations of heterotaxy patients have been reviewed by Rubin(29), and Gilljam (30).

In summary, the present report highlights the high incidence of RHD, the growing incidence of ACHD, and the specifics of situs topography. The unusual surgical approach to this combined condition is described.

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