secondary either to the direct effect of etoposide, previous mediastinal irradiation or both.

In conclusion, this patient suffered a myocardial infarction following combination chemotherapy, and although most of the literary evidence points to cisplatin and vincristine as the cardiovasculatory toxic agents, the role of etoposide must not be overlooked, particularly as this agent is frequently used in young people with potentially curable neoplasms.

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References


[7] X-ray confirmed dextrocardia with a cardio-thoracic ratio of 0.6. Electrocardiogram showed atrial fibrillation with frequent unifocal ventricular ectopics. On echocardiography, the systemic ventricle was anterior with poor contraction but the performance of the pulmonary ventricle was satisfactory. There was no significant incompetence of the heart valves and no intracardiac shunts. Cardiac catheterization demonstrated normal coronary arteries, and a satisfactory coarctation repair. The pulmonary artery capillary wedge pressure was elevated, and the calculated cardiac output low (Table 1). Metabolic exercise testing showed a maximum oxygen uptake of 51% of the predicted value. Resting ejection fraction measured by multigated isotope scanning was 24%.

The patient was considered for transplantation by our Transplant Service, but declined in view of the likely technical difficulties involved. She elected to undergo cardiomyoplasty. At operation it was confirmed that the morphologically left ventricle was placed anteriorly with the apex to the right, and the morphologically right ventricle posterior and to the left. Cardiomyoplasty was performed, using the right latissimus dorsi muscle in an antero-posteriorly orientated wrap, whereby the proximal and most powerful part of this muscle was used to cover the systemic ventricle. Muscle stimulating electrodes and a ventricular sensing electrode were placed and connected to a cardiomyostimulator (Medtronic SP1005). Postoperative recovery was uneventful. A 6-week muscle transformation programme was commenced 2 weeks following surgery. Following completion of transformation the muscle was stimulated to provide 1:2 (muscle:heart) systolic support.

The patient has remained well to 17 months follow-up, with a marked relief of symptoms and she is now in New York Heart Association class I. Haemodynamic measurements at one year follow-up showed a slight elevation in pulmonary artery wedge pressure, and unchanged ejection fraction. Resting cardiac output by thermodilution technique was elevated by 35% and there was a substantial improvement in the maximum oxygen uptake on exercise.

Although cardiac transplantation in dextrocardia has been described, it may be technically demanding. Cardiomyoplasty may be an attractive option in selected patients and when the right-sided latissimus dorsi muscle is used, it has the additional advantage that this muscle is usually stronger and larger than its counterpart in a right-handed person. To our knowledge, this is the first report on cardiomyoplasty in a patient with dextrocardia.

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Table 1 Haemodynamic parameters

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Before operation</th>
<th>1 year follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>PA pressure (mmHg)</td>
<td>48/16, Mean 28</td>
<td>48/20, Mean 31</td>
</tr>
<tr>
<td>PAWP (mmHg)</td>
<td>16</td>
<td>17</td>
</tr>
<tr>
<td>CO (l.min⁻¹)</td>
<td>3.4</td>
<td>4.6</td>
</tr>
<tr>
<td>Resting EF (%)</td>
<td>24</td>
<td>24</td>
</tr>
<tr>
<td>VO₂ max (ml.kg⁻¹.min⁻¹)</td>
<td>12.85</td>
<td>15.54</td>
</tr>
<tr>
<td>Heart rate</td>
<td>80</td>
<td>80</td>
</tr>
</tbody>
</table>

CO=cardiac output; EF=ejection fraction; PA=pulmonary artery; PAWP=pulmonary artery wedge pressure; VO₂ max=maximum oxygen uptake.
Pulmonary valve stenosis in a patient with ataxia telangiectasia

We have followed up a patient affected by ataxia telangiectasia, who at the age of 23 developed the signs of pulmonary valve stenosis, an association not previously reported to our knowledge.

The patient, a 24-year-old female, was the first born of healthy, blood-related parents. Her family history was unremarkable. Pregnancy, birth and post-partum development were normal until the age of 18 months, when truncal ataxia and ataxia of gait became apparent. These defects worsened and ataxia was subsequently noted in the arms, to be followed by nystagmus, myoclonic jerks, titubation, dystrophic speech, hyporeflexia, tremor and choreoathetosis.

Telangiectasies were first noted in the conjunctiva, at age 3 years, and these later developed in the skin. Upper respiratory infections and pneumonia had been frequent during the patient's infancy and childhood. At 16 years of age, by which time she was confined to a wheelchair, symptoms of diabetes mellitus appeared. At 23 years of age the patient was referred to our Institution because of fatigue, dyspnoea on exertion and tachycardia. On physical examination, her skin was pale and dry with evidence of acrocyanosis and acral and cutaneous telangiectasies. She had a dull, flat facies, cervicosternal kipscholiosis, equinovarus deformity of the feet and amyotrophy of distal limbs. Cardiac auscultation revealed a holosystolic rough ejection murmur (3/6 Levine) in the second left interspace, radiating to the supraclavian area. The patient showed evidence of a severe cerebellar syndrome with choreoathetoid movements and generalized dystonia. Mental retardation was severe. Complete physical examination revealed no other abnormalities. A chest radiograph showed a prominent main pulmonary artery segment. Electrocardiography showed pulmonary P waves in leads D1, D2, AVF and V1, a normal P-Q interval and right axis deviation in the frontal plane (+110°). R waves were 20 mm leads V1, V2 and aVR. Two-dimensional echocardiography demonstrated thickening of the pulmonary valve and right ventricular hypertrophy; Doppler study revealed a gradient across the pulmonary valve (Fig. 1). ECG and echocardiography were performed in all family members and results were unremarkable. The patients' serum alpha-fetoprotein and carcino embryonic antigen were five times normal; the IgA level was 14.5 mg dl⁻¹ (normal 90-450 mg dl⁻¹). A magnetic resonance image of the brain revealed severe atrophy of the cerebellar vermis and hemispheres.

Ataxia telangiectasia is an autosomal recessive multisystem disorder characterized clinically by progressive cerebellar ataxia, ocular telangiectasies, and variable immunodeficiency with recurrent sinopulmonary infections[1]. The gene responsible, ATM (ataxia telangiectasia, mutated) has been identified by positional cloning on chromosome 11q22-23[2].

Heterozygous carriers of the ataxia telangiectasia gene, estimated to comprise about 1-3% of the general population, have a two-to-six elevated risk of dying from cancer and a predisposition to ischaemic heart disease[3]. However, there is still not enough information in the literature about the incidence of cardiac anomalies in homozygotes with the ataxia telangiectasia syndrome[4,5]. Bastianon and Chessi[6] reported clinical and echocardiographic data for a series of 12 patients with proven ataxia telangiectasia, showing that mitral valve prolapse was present in five patients and aortic root dilatation in one.

Pulmonary valve stenosis, found at a frequency of 5-7% among all cardiac anomalies, has been reported to have a familial incidence, with a recurrence rate of 2-4%, and to be frequently associated with several chromosomal and malformative syndromes, and heritable disorders of connective tissue[5]. Patients with ataxia telangiectasia have abnormalities of collagen with decreased fibronectin mRNA in fibroblasts[7].

The present case of pulmonary valve stenosis could be an isolated lesion coincidentally associated with ataxia telangiectasia, but since genetic factors may play a role in congenital heart disease, in particular in patients with a defect in a single gene, such a cardiac anomaly could perhaps be regarded as part of the syndrome.

The patients died at age 24 years, of gastrointestinal cancer complications.

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References


Sick sinus syndrome in association with malignant lymphoma

Metastasis of malignant lymphoma to the heart is an uncommon feature of...