J.D. Milton
R.H.T. Edwards
G.B. Clements¹

Department of Medicine,
University of Liverpool,
Liverpool L69 3BX; and
¹Regional Virus Laboratory,
Ruchill Hospital,
Glasgow, G20 9NB, UK.

References

1. A report – chronic fatigue syndrome: guidelines for research. *J R Soc Med* 1991, **84**: 118–121.
2. Cunningham, L.C., Bowles, N.E., Lane, R.M.J., Dubowitz, V. & Archard, L.C. Persistence of enteroviral RNA in chronic fatigue syndrome is associated with the abnormal production of equal amounts of positive and negative strands of enteroviral RNA. *J Gen Virol* 1990, **71**: 1399–1402.
3. Morrison, L.J.A., Behan, W.M.H. & Behan, P.O. Changes in natural killer cell phenotype in patients with postviral fatigue syndrome. *Clin Exp Immunol* 1991, **83**: 441–446.

Myotonia dystrophica – first presentation as severe left ventricular failure complicating dilated cardiomyopathy

Sir,

Myotonia dystrophica is an autosomal dominant multi-system disorder which in adults affects the specialized cardiac conduction tissue frequently, but the myocardium itself only rarely clinically,¹ although electron microscopic studies show early myocardial involvement.² Cardiomyopathy has been described,^{3,4} but we are unaware of reports of first presentation of myotonic dystrophy with dilated cardiomyopathy and left ventricular failure.

A previously well, 30 year old infertile man presented with breathlessness at rest and paroxysmal nocturnal dyspnoea. There was no significant family or drug history. He was orthopnoeic and hypotensive (blood pressure 90/70 mmHg) with a raised jugular venous pressure. His pulse was 112/min and regular. There was a prominent left ventricular third heart sound, accentuated pulmonary second sound and a soft systolic murmur in the mitral area. He was also noted to have frontal baldness, drooping eyelids, wasting of the masseters, myotonia, small firm testes, and lenticular opacities on slit lamp examination. There was no significant muscle weakness.

Electrocardiography showed a sinus tachycardia, complete left bundle branch block and left anterior hemiblock. Chest X-ray confirmed left ventricular failure. Echocardiography revealed a moderately dilated left ventricle, hypokinesia of the interventricular septum and left ventricular posterior wall, E point septal separation of 25 mm and a reduced aortic valve opening of 1.2 cm. These parameters were suggestive of moderate to severe left ventricular dysfunction complicating dilated cardiomyopathy.

Dilated cardiomyopathy is rare in adults with myotonia dystrophica. Why this should be so is unknown, as cardiac conduction tissue, which is commonly involved, both sub-clinically and clinically, has a close embryologic

link with cardiac muscle. Perhaps the susceptibility of the myocardium to the biochemical defect affecting conduction tissue is determined by hitherto unknown genetic or environmental risk factors. It is also uncommon for patients with myotonic dystrophy to present with severe left ventricular failure as our patient did. Cardiac muscle involvement is commonly occult throughout life and there is no consistent correlation with skeletal muscle involvement.¹ Electrocardiological and electrophysiological studies show abnormal sinus node function in about 20% and His-Purkinje disease in 80% of patients.¹ Isolated left bundle branch block and left anterior hemiblock may occur in 13–24% of patients.⁵ But it is rare for a combination of left anterior hemiblock and complete left bundle branch block to occur in the same patient. This may have been a marker of severe cardiac muscle involvement.

L.D.K.E. Premawardhana[†]

G. Thirunavakarasu
Department of Medicine/Cardiology,
King Khaled General Hospital,
Al Kharj, Riyadh,
Saudi Arabia.

[†]Department of Medicine,
University Hospital of Wales,
Heath Park,
Cardiff CF4 4XN.

References

1. Perloff, J.K., Stevenson, W.G., Roberts, N.K., Cabeen, W. & Weiss, J. Cardiac involvement in myotonia dystrophica; a prospective study of 25 patients. *Am J Cardiol* 1984, **54**: 1074–1081.
2. Motta, J., Guilleminault, C., Billingham, M., Barry, W. & Mason, J. Cardiac abnormalities in myotonic dystrophy. Electrophysiologic and histopathologic studies. *Am J Med* 1979, **67**: 467–473.
3. Pandullo, C., Nicolosi, G.L. & Scardi, S. Hypertrophic cardiomyopathy associated with myotonic dystrophy. *Int J Cardiol* 1987, **16**: 205–208.
4. Lin, A.E., Mitchell, F.M., Fitz, R.W. & Doyle, J.J. Dilated cardiomyopathy in myotonic dystrophy. *J Am Coll Cardiol* 1989, **13**: 262–263.
5. Komajda, M., Frank, R., Vedel, J., Fontaine, G., Petitot, J. & Groagoeat, Y. Intracardiac conduction defects in dystrophica myotonica. Electrophysiological study of 12 cases. *Br Heart J* 1980, **43**: 315–320.

Intussusception in ileostomy in a pregnant woman

Sir,

Intussusception in stomata is rare^{1–3} and only two cases have been reported previously in the literature reviewed since 1950.^{4,5} A case of intussusception of ileostomy in a woman pregnant with twins is presented and its mechanism described.

A 31 year old woman with ileostomy presented with 'prolapse' of her stoma, associated with colicky abdominal pain. She was 21 weeks pregnant with twins. Abdominal examination showed a uterus compatible with her date of pregnancy, multiple scars, no area of tenderness and normal bowel sounds. Her haematological investigations were within normal limits.