

# Letters

## LOBAR PNEUMONIA TREATED BY MUSGRAVE PARK PHYSICIANS.

Editor,

In the excellent historical article by John Hedley-Whyte<sup>1</sup>, I saw the photograph of Sir Alexander Fleming with Professor William Thomson on the doorsteps of Number 25 University Square and I remembered that in my grandfathers visitors book there were the signatures of Sir Alexander and Lady Fleming not only at number 12 University square (figure 1), but also at Greenlawn in Donaghadee (figure 2). The exact date is not clear but I suspect about April 1942.



Fig 1. Visitors book from 12 University Square.

Professor CG Lowry (known as CG) and Professor Thomson (known as WWD) were close friends, colleagues and neighbours both in University Square, CG at number 12, and WWD at Number 25, and also next door at Donaghadee, and hence this accounts for the above records of those events.

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## CORONARY ARTERY DISEASE: ANATOMY AND PRESENTATION IN IDENTICAL TWINS

Editor,

A 47 year old man (twin 1) was admitted electively for coronary angiography following an acute myocardial



Fig 2. Visitors book from Greenlawn, Donaghadee

infarction (MI) one month previously. His risk factor profile included smoking, a positive family history, hypertension and hypercholesterolaemia. On the day of admission, it was discovered that his identical twin brother (twin 2) was an elective inpatient for coronary angiography. His history included acute MI aged 42 years, with subsequent percutaneous coronary intervention to the circumflex. His risk factor profile included previous MI, a positive family history, hypertension and hypercholesterolaemia.

Coronary angiograms were performed on consecutive days. Coronary arterial anatomy was discordant between the twins. Angiographic images from twin 1 are shown in figure 1 (panels 1a-1c), beside matched images from twin 2 (panels 2a-2c). In twin 1 the left main stem bifurcates into left anterior descending (LAD) and circumflex (CX) branches (panel 1a), while in twin 2 it trifurcates into an LAD, CX and ramus intermedius branch (panel 2a). The first obtuse marginal branch (OM1) arises and bifurcates proximally in twin 1 (panels 1a and 1b) but arises and bifurcates more distally in twin 2 (panels 2a and 2b). The right coronary artery supplies a prominent sinus node branch in twin 1 (SA node, panel 1c) which is not apparent in twin 2 (panel 2c).

Coronary artery disease distribution was also discordant between the twins. Twin 1 was found to have a normal left main stem, with a long area of moderate to severe disease in the mid part of the LAD. A large diagonal branch had a 90% ostial lesion. There was a 50% lesion in the main CX and a 90% lesion in its first marginal branch. The right coronary artery was diffusely diseased. Twin 2 had a normal left main stem, with an angiographically near-normal LAD. The CX was diffusely diseased. The right coronary artery was diffusely diseased but with no significant stenosis.

Our observation of discordant coronary artery distribution and coronary atherosclerosis in identical twins supports the findings of previous observational studies<sup>1,2</sup>. Furthermore age at first cardiac event, type of cardiac event and risk factor profile show concordance in this pair of identical twins, also consistent with previous observations<sup>2</sup>.

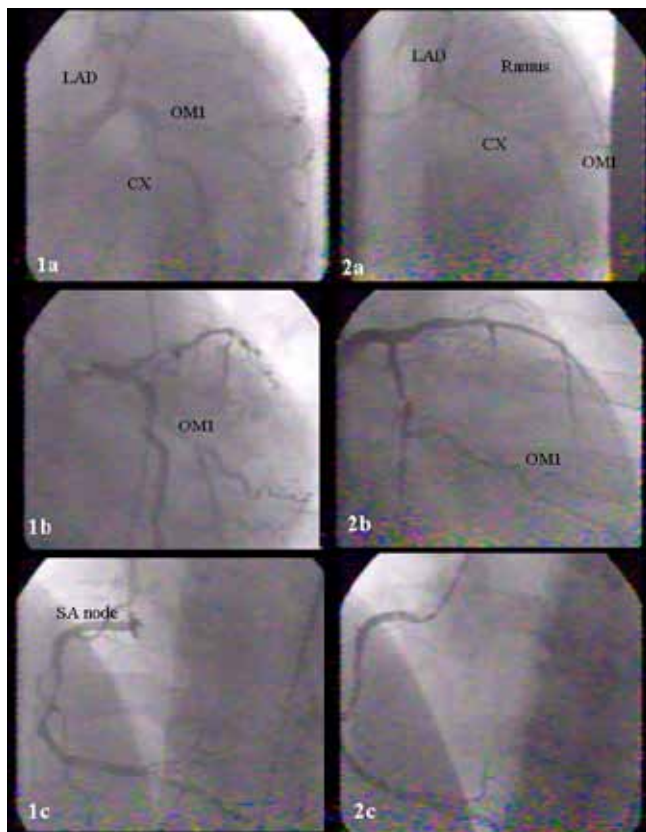


Fig 1. Coronary angiograms from twin I ( left panel, 1a-c) and twin 2 (Right panel, 2a-c)

We conclude that coronary anatomy is independent of the human genome. Disease lesion sites are at least partly independent of the human genome. In contrast, age at first cardiac event, type of cardiac event and risk factor profile appear to be more closely related to genetic profile. We suggest that when one twin presents with IHD, the second should be subject to increased medical surveillance

The authors have no conflict of interest

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#### DRUGS, ELECTROLYTES AND TAKO-TSUBO CARDIOMYOPATHY: TRIPLE AETIOLOGY OF ACQUIRED LONG QT SYNDROME AND TORSADES DE POINTES.

Editor,

Physical or emotional stress can have unforeseen consequences. We document a 67 year-old female admitted with syncope following emotional stress. She had a history of depression and had been “crying and crying all day”. In addition, she had a history of ileostomy following severe diverticular disease. Her daily medication included ondansetron 4mg b.d. for nausea and fluoxetine 60mg for depression.

On admission, serum magnesium was low at 0.73mmol/l (0.75 – 1.25) and serum potassium was 3.9mmol/l (3.5 – 5.1). Troponin I was mildly elevated at 0.17u/l (0 – 0.04). B-type natriuretic peptide (BNP, Abbott) was grossly elevated at 2569pg/ml (normal < 100). Initial ECG (fig 1) showed new T wave inversion in ECG leads; II, III, aVf and V1 through to V6 with a prolonged corrected QT interval (QTc) of 524ms (upper limit of normal for females = 450ms). An ECG dated June 2007 was normal apart from a QTc of 509ms. She was initially treated as an anterior non-ST segment elevation myocardial infarction. Shortly after admission, she developed polymorphic ventricular tachycardia (torsades de pointes, figure 2). The risk of torsades de pointes increases substantially once QTc is > 500 ms. This was treated with a 200J DC shock, 4mmol of intravenous magnesium with oral beta-blocker, and potassium therapy. Further self-terminating runs of torsades de pointes occurred when her potassium levels dipped below 4mmol/l.

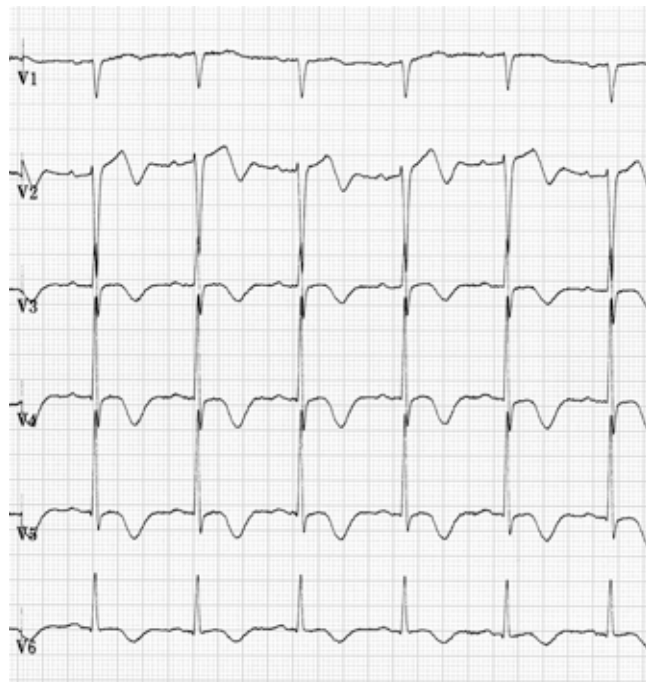


Fig 1. Leads V1 – V6 of admission 12-lead ECG showing T wave inversion resembling non-ST segment elevation MI. QTc is greatly prolonged at 524ms.

On day two, she underwent cardiac catheterisation, which showed normal coronary arteries but marked impairment of systolic function in the apical half of the left ventricle with a characteristic “ballooning” appearance (figure 3). These findings, in association with physical or emotional strain, are diagnostic of tako-tsubo cardiomyopathy. Oral magnesium supplements and bisoprolol 5mg were added in to her medication. Ondansetron and fluoxetine both prolong the QT interval and were stopped. A cardio-defibrillator device was

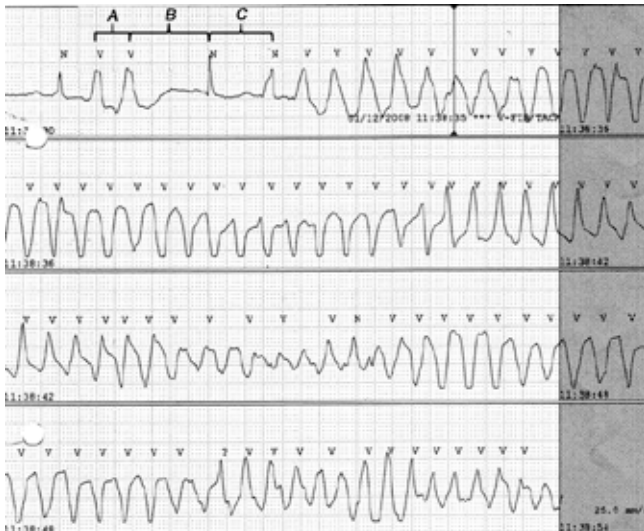


Fig 2. ECG monitor tracing shows a characteristic short (A), long (B), short (C) sequence of ventricular activity initiating polymorphic ventricular tachycardia also known as "torsades de pointes" for its twisting QRS axis about the iso-electric line.

implanted due to continued risk of arrhythmia from electrolyte loss from the ileostomy. The QTc came down to 454ms and BNP fell to 179pg/ml at discharge.

QT prolongation is the surface ECG manifestation of abnormal repolarisation of myocardial cells due to problems with cellular ion channels. The disorder is classified as either congenital or acquired. Acquired QT prolongation may be due to:

1. Electrolyte depletion, particularly potassium or magnesium,
2. Drugs that affect myocardial ion channels
3. A feature of tako-tsubo cardiomyopathy, a catecholamine induced metabolic disorder of myocardial cells caused by physical or emotional stress, especially seen in older females<sup>1,2</sup>.

A reference list of drugs causing QT prolongation is available from the University of Arizona (<http://www.azcert.org>) or the British National Formulary.

Initial presentation and ECGs in tako-tsubo cardiomyopathy are similar to an anterior ST or non-ST segment myocardial infarction but often with QT prolongation. A small troponin rise may be seen but coronary arteries are normal with a characteristic "apical ballooning" or Japanese octopus pot ("tako-tsubo") pattern seen on ventriculography. Beta-blockade is a key element of treatment. The ventricular changes are mostly reversible if the patient survives the acute phase<sup>3</sup>.

Our patient had all three causes of an acquired QT prolongation - excessive secretion from her ileostomy producing hypomagnesaemia, daily ondansetron and fluoxetine therapy, and acute tako-tsubo cardiomyopathy. We believe the development of tako-tsubo cardiomyopathy exacerbated our patient's pre-existing QT prolongation to a degree where potentially fatal arrhythmias occurred.

A case of congenital long QT syndrome and tako-tsubo cardiomyopathy with torsades de pointes has been described<sup>4</sup> but MEDLINE and PubMed searching (keywords: long QT

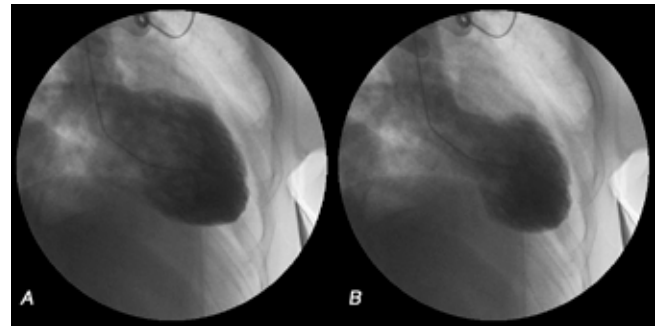


Fig 3. Left ventriculography during diastole (A) and systole (B) demonstrate typical left ventricular apical ballooning and hyper-contractile base of tako-tsubo cardiomyopathy.

and cardiomyopathy) revealed no acquired cases. Tako-tsubo cardiomyopathy induced by physical or emotional stress may exacerbate an underlying long QT syndrome with risk of sudden cardiac death.

The authors have no conflict of interest

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#### PSEUDOMYXOMA PERITONEI PRESENTING AS INGUINAL HERNIA.

Editor,

Pseudomyxoma peritonei (PMP) is an uncommon disease with varied presentations. We present two cases presenting at inguinal hernia repair.

**Case 1:** A 41 year-old man presented for right inguinal hernia repair. An encysted swelling was discovered at surgery. Histopathology of the sac showed chronic inflammatory tissue containing lakes of mucin but no neoplastic epithelial cells. Postoperative CT scan showed thickening around the caecum with a fluid collection and abnormality related to the appendix. Colonoscopy and biopsies were normal. The patient was referred to the National Specialist Commissioning Advisory Group Pseudomyxoma Peritonei Centre (Basingstoke) where a laparotomy revealed a

perforated appendiceal tumour and widespread peritoneal disease. A radical greater omentectomy, right hemicolectomy, cholecystectomy and removal of peritoneal disease was performed. Intraperitoneal chemotherapy was administered and the patient made a satisfactory recovery.

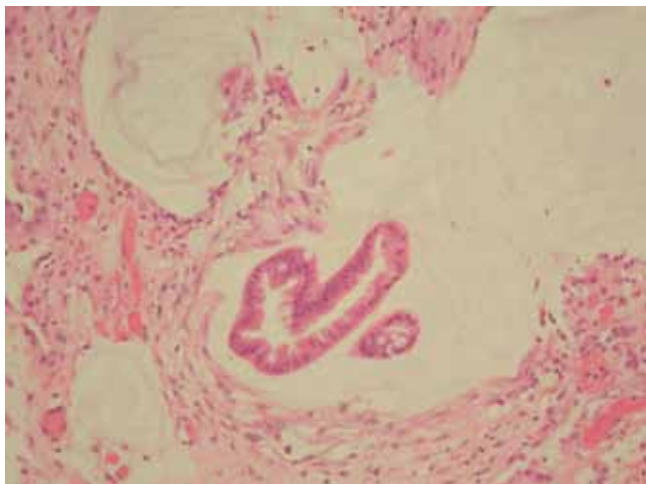


Fig 1. H&E stain (x200)

**Case 2:** A 73 year-old man presented for right inguinal hernia repair. At surgery the hernial sac appeared thickened. Histopathology (fig 1) showed a thick inner wall composed of chronic inflammatory tissue containing lakes of mucin and well-differentiated enteric type glandular epithelium with minimal cytonuclear atypia. This was considered diagnostic of PMP. Immunohistochemistry showed cytokeratin 7-/20+ staining, characteristic of pseudomyxoma peritonei of large bowel, especially appendiceal origin (fig 2). Post-operative CT scan showed omental cake and ascites. The appendix appeared normal. CEA was raised at 45ng/ml; Ca19-9 was normal. Due to the extent of disease the patient was managed conservatively with follow up imaging and monitoring of tumour markers.

**Discussion:** PMP is characterised by the build up of mucoid material and fluid within the abdomen and pelvis. The diagnosis is challenging due to the range of presenting features. Patients typically present with abdominal pain, increased abdominal girth or an abdominal mass. A recent review of the clinical presentation of PMP found new onset hernia to be the fourth commonest presentation (14% of cases)<sup>1</sup>.

It is well established that the majority of cases are of appendiceal origin<sup>2</sup>. The ovary is rarely the origin of PMP except for the rare case of an intestinal type mucinous neoplasm arising in a teratoma<sup>3</sup>. The ovary may however be a site of secondary spread from the appendix. There are two variants of PMP; Disseminated peritoneal adenomucinosis (DPAM) and peritoneal mucinous carcinomatosis (PMCA). DPAM arises from an appendiceal mucinous adenoma and peritoneal mucinous carcinomatosis (PMCA) is associated with mucinous gastrointestinal adenocarcinomas<sup>4</sup>. The CK7-/CK20+ immunohistochemical staining pattern is characteristic of pseudomyxoma peritonei of gastrointestinal, especially appendiceal origin (Case 2). Primary ovarian mucinous tumours are characteristically CK7+/CK20-<sup>3</sup>.



Fig 2. Cytokeratin 20 (x400)

If mucoid material or fluid is found at the time of hernia repair it should be sent to histopathology and the hernia repaired without mesh, thus avoiding trapping tumour cells. Tumour markers should be sent, a CT scan arranged, and the patient referred to a specialist treatment centre once the diagnosis is confirmed<sup>5</sup>. Treatment consists of a combination of peritonectomy procedures and intraperitoneal chemotherapy. This approach has reported 5-year survival rates in excess of 80%<sup>5</sup>.

**Conclusion:** These cases emphasise the importance of considering PMP if a thickened sac or mucinous material is encountered at hernia repair.

The authors have no conflict of interest.

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