

CASE REPORTS

Phrenic nerve injury following blunt trauma

David Bell, Ajith Siriwardena

Abstract

Phrenic nerve trauma in the absence of direct injury is unusual and may present diagnostic difficulty. Diaphragmatic paralysis resulting from phrenic nerve injury may closely mimic diaphragmatic rupture. This case highlights the value of magnetic resonance imaging in establishing diaphragmatic integrity and of ultrasonographic assessment during respiratory excursion in confirming diaphragmatic paralysis. In cases of non-contact injury involving torsional injury to the neck, an index of clinical awareness may help to establish the diagnosis of phrenic nerve trauma.

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Keywords: phrenic nerve injury; blunt trauma

Case report

A 36 year old man was admitted to the accident and emergency department two hours after a road traffic accident. The patient was driving a car that had been struck on the near-side by another vehicle. The patient’s vehicle had been stationary at the time of impact. The patient was wearing a seatbelt and reported that he had not struck his head. There was no history of loss of consciousness. He complained of pain in the head, neck and back. He had also briefly experienced paraesthesiae in the right hand but this had resolved by the time of arrival in the department.

At primary survey his neck was immobilised in a hard collar. He was maintaining his airway, self ventilating with a respiratory rate of 20/minute and there was decreased air entry to the right lower zone. The trachea was central. Cardiovascular examination was normal. Oxygen saturation was 97% on air and an electrocardiograph was normal. Secondary survey demonstrated severe lumbar spine tenderness over T12 and L1 vertebrae but no obvious neurological deficit. Physical examination was otherwise normal. His past medical history was significant for a myocardial infarction one year previously.

Radiographs of the cervical spine were normal. Lumbar spine films demonstrated a stable anterior wedge compression fracture of T12. Chest radiograph showed an elevated right hemi-diaphragm (fig 1). This was not evident on a chest film taken 12 months previously (fig 2). A provisional diagnosis of



Figure 1 Posteroanterior chest radiograph taken after injury showing elevated right hemi-diaphragm.

diaphragmatic rupture was made. Computed tomography demonstrated mild rotation of the axis of the liver (anti-clockwise rotation through the plane of the middle hepatic vein) compatible with diaphragmatic rupture. However, a magnetic resonance scan confirmed that the diaphragm was intact but elevated. Ultrasound scan with respiratory excursion demonstrated paralysis of the right hemi-diaphragm.

A diagnosis of right hemi-diaphragmatic paralysis secondary to phrenic nerve damage



Figure 2 Posteroanterior chest radiograph taken 12 months before injury showing normal position of right hemi-diaphragm.

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was made. The injury was treated conservatively with non-opioid analgesia for the lumbar spine injury. The patient remains well six months after injury with no clinical evidence of respiratory compromise. He has declined further assessment of diaphragmatic function.

Discussion

Traumatic phrenic nerve injury is well recognised after both penetrating and blunt trauma to the neck.^{1 2} In contrast, injury as a result of distraction or stretching of the nerve is rare.³ In several of these previous reports, a component of nerve damage may have been as a result of blunt trauma. There was no evidence of blunt trauma in this case with the mechanism of injury thought to be lateral hyperextension of the neck. There are no previous reports of phrenic nerve palsy by this mechanism.

Clinical manifestations of this injury include breathlessness, orthopnoea and respiratory distress.⁴ The diagnosis may be suspected on chest radiography and computed tomography

and confirmed by fluoroscopy or ultrasonography with respiratory excursion. An important practical consideration is that the clinical findings and radiological appearances on plain radiographs and computed tomography may mimic diaphragmatic rupture.

In summary, this case highlights a rare cause of phrenic nerve injury in the absence of direct trauma. The clinical presentation may closely resemble diaphragmatic rupture.

Contributors

David Bell initiated the writing of the report and the MEDLINE search. Ajith Siriwardena supervised the writing of the report and the phrasing of the final draft and reviewed the adequacy of the literature search and review of relevant publications.

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Conflicts of interest: none.

- 1 Iverson LJ, Mittal A, Dugan DJ, *et al.* Injuries to the phrenic nerve resulting in diaphragmatic paralysis with special reference to stretch trauma. *Am J Surg* 1976;132:263–9.
- 2 Dalshaug GB, Rothwell BC. Diaphragmatic paralysis following minor blunt trauma. *Journal of Trauma, Injury, Infection and Critical Care* 1999;47:413–15.
- 3 Snyder RW, Kukora SJ, Bothwell WN, *et al.* Phrenic nerve injury following stretch trauma. *J Trauma* 1994;36:734–6.
- 4 Carter RE. Unilateral diaphragmatic paralysis in spinal cord injury. *Paraplegia* 1980;18:267–74.

Non-penetrating chest blows and sudden death in the young

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Abstract

Sudden death in the young after low energy anterior chest wall impact is an under-recognised phenomenon in this country. Review of the literature yields several American references to commotio cordis, mainly in the context of sporting events. Two cases are reported of sudden death in young men as a result of blunt impact anterior chest wall trauma. It is suggested that these cases draw attention to a lethal condition of which many practitioners are unaware.

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Keywords: chest blows; sudden death

Case report 1

The deceased was a 15 year old boy who collapsed to the ground immediately after being struck centrally in the chest with a 500 g stone, thrown from a distance of 8 to 10 feet during a gang fight. Upon collapse he was said to have suffered a fit and was carried bodily a short distance by friends and laid on the ground. There was no basic life support until ambulance paramedics arrived, five minutes after receiving the emergency call.

The cardiac rhythm at the scene was pulseless electrical activity, which degenerated to ventricular fibrillation resistant to electrical cardioversion. Appropriate advanced life support continued and he arrived at the accident and emergency (A&E) department 27 minutes after the emergency call. His pupils were fixed and dilated and he had an agonal rhythm, interrupted by runs of ventricular tachycardia and episodes of ventricular fibrillation. Right needle thoracocentesis was performed because of reduced air entry on auscultation but no air was aspirated. Internal jugular and femoral venous lines were inserted and a crystalloid bolus was infused. Ventricular fibrillation developed, but electrical cardioversion proved unsuccessful. His rhythm degenerated into asystole and resuscitation attempts were stopped.

NECROPSY FINDINGS

There were signs of medical intervention in the form of needle puncture marks in the cubital fossae, right lower neck, right groin and right upper anterior chest, and defibrillator paddle marks on the anterior chest wall. There were minor abrasions to both anterior knees, the fingers of the right hand and the left eyebrow, cheek, and chin. There was no significant externally apparent injury to the anterior chest wall. There was minor bruising over the left pectoralis major muscle. Internal examination revealed moderate bruising of the lingula and patchy subpleural bruising on the posterior

right and left lower lung lobes. There was a 15 mm long and 8 mm deep laceration within the left ventricular myocardium at the apex. There was no natural disease and toxicological analyses were negative. Death was attributed to blunt force chest trauma.

Case report 2

A 19 year old man was playing in goal in a five a side football game when he was struck in the central chest by a leather football, kicked with considerable force from a range of only a few yards. Although initially in obvious discomfort and “winded”, he continued to stand in goal for an estimated 5–10 minutes before crying out and collapsing. Bystander cardiopulmonary resuscitation was started promptly by a doctor participating in a neighbouring game. Ambulance paramedics performed advanced life support, including repeated administration of epinephrine and attempts at electrical cardioversion. On arrival in the A&E department approximately 70 minutes after the collapse he was in asystole. Further attempts at resuscitation, including pericardiocentesis and external pacing, were performed but unfortunately failed to achieve return of spontaneous circulation.

NECROPSY FINDINGS

There were signs of medical intervention in the form of electrode pads, endotracheal intubation and a needle puncture mark in the left cubital fossa. Marked upper anterior mediastinal bruising was present in relation to right subclavian cannulation. There was 350 ml of blood within the pericardial sac and a small puncture mark over the lower sternum entering the right ventricular apex, representing the attempt at pericardiocentesis. There was no other myocardial abnormality and no injury to the chest wall. There was no natural disease but toxicological analyses were not performed. Death was attributed to blunt force chest trauma resulting from being struck on the chest by the football.

Discussion

We suggest that the above cases illustrate death attributable to primary arrhythmia occurring after blunt chest trauma. This is often described as commotio cordis, which typically occurs in the absence of structural cardiac damage. Collapse is usually near instantaneous, but there can be a period of activity following impact, usually only of a few seconds duration.^{1–3}

Most previous reports are concentrated in American literature and occur mainly in baseball and ice hockey, where projectile impacts to the precordium are frequent.^{1–3} Such projectiles cause a focused impact of short duration

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with virtually no rebound, allowing almost complete transfer of kinetic energy to the chest wall.³ Commotio cordis has not been reported in the game of cricket, Britain's nearest sporting equivalent. Maron *et al*⁴ have described four cases of commotio cordis occurring in relation to criminal activity.

An experimental model for commotio cordis suggests that the impact of projectiles on the chest wall of pigs during the period of cardiac repolarisation, prior to the peak of the T wave, typically induced ventricular fibrillation.⁵ This situation probably corresponds to the typical presentation reported in most fatalities, causing immediate collapse and cardiac arrest. In contrast, impact during the QRS complex is more likely to induce complete heart block, ST segment elevation or left bundle branch block.⁵ Other reported ECG abnormalities include sinoatrial nodal dysfunction, atrial fibrillation, right bundle branch block and ventricular extrasystoles.^{6,7}

In the first case, the sequence of events after impact were typical of commotio cordis. However, it is unusual in that it occurred in a homicidal manner, with an object that is heavier than usual, thrown over a shorter distance and at lower velocity. In this case there was evidence of structural cardiac damage. The apical endocardial laceration may be attributable to the hydraulic ram effect of ventricular compression, causing blood to be forced into the blind-ending apex of the left ventricle. This could have been attributable either to impact by the stone itself or may represent resuscitation artefact.⁸

Our second reported case differed significantly from the usual presentation of commotio cordis in that collapse occurred several minutes after chest impact. Blood was found in the pericardial sac at necropsy suggesting a slowly developing cardiac tamponade may have been

responsible for death. The attempt at pericardiocentesis produced no blood, suggesting that there was no sizeable collection of blood at the time of resuscitation. This reflects the difficulty in determining whether the pericardial haemorrhage resulted from an injury at the time of trauma or by attempts at pericardiocentesis. We considered it possible that chest impact, presumably during the QRS complex, produced one of the above electrical abnormalities that then degenerated into a malignant arrhythmia with cardiac arrest after some minutes.

The severity of impact required to cause death after a blow to the chest is often unremarkable and careful enquiry should be made for a history of chest wall impact after cardiac arrest or sudden death in the young. We further suggest that the classic impression of instantaneous collapse may not be true in every case.

Contributors

Shobhan Thakore, Emily Rogena, Zhang Peng, David Sadler and Michael Johnston collected the clinical data on the two cases and wrote the paper. David Sadler and Michael Johnston act as guarantors of the paper.

- 1 Abrunzo TJ. Commotio cordis—the single, most common cause of traumatic death in youth baseball. *American Journal of Diseases of Children* 1991;145:1279–82.
- 2 Maron BJ, Strasburger JF, Kugler JD, *et al*. Survival following blunt chest impact-induced cardiac arrest during sports activities in young athletes. *Am J Cardiol* 1997;79:840–1.
- 3 Maron BJ, Poliac LC, Kaplan JA, *et al*. Blunt impact to the chest leading to sudden-death from cardiac-arrest during sports activities. *N Engl J Med* 1995;333:337–42.
- 4 Maron BJ, Link MS, Wang PJ, *et al*. Clinical profile of commotio cordis: an under appreciated cause of sudden death in the young during sports and other activities. *J Cardiovasc Electrophysiol* 1999;10:114–20.
- 5 Link MS, Wang PJ, Pandian NG, *et al*. An experimental model of sudden death due to low-energy chest-wall impact (commotio cordis). *N Engl J Med* 1998;338:1805–11.
- 6 Crown LA, Hawkins W. Commotio cordis: clinical implications of blunt cardiac trauma. *Am Fam Physician* 1997;55:2467–70.
- 7 Kumpuris AG, Casale TB, Mokotoff DM, *et al*. Right bundle branch block; occurrence following nonpenetrating chest trauma without evidence of cardiac contusion. *JAMA* 1979;242:172–3.
- 8 Leadbeater S, Knight B. Resuscitation artefact. *Med, Sci Law* 1988;23:200–4.

Extracorporeal rewarming in a severely hypothermic patient using venovenous haemofiltration in the accident and emergency department

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Abstract

Severe hypothermia is a medical emergency and requires active and occasionally rapid core rewarming to prevent cardiac arrhythmias and death. In the accident and emergency department rewarming is often limited to warmed intravenous fluids, heated blankets, gastric and bladder lavage. Extracorporeal methods, which rewarm core blood directly, for example haemodialysis and cardiopulmonary bypass, require expertise and equipment not always found in a district

general hospital. Venovenous haemofiltration is now commonly found in district general hospitals around the country and can be used safely for core rewarming. A case is reported of a severely hypothermic elderly patient successfully rewarmed using venovenous haemofiltration, in an accident and emergency department, when other conventional methods had failed.

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Hypothermia is defined as a core body temperature below 35°C.¹ Patients presenting with severe hypothermia (<28°C)² are at high risk of serious arrhythmias and asystole.¹

Rewarming severely hypothermic patients in the accident and emergency (A&E) department initially requires non-invasive methods, which are usually successful. However, if these methods fail, or the patient is in circulatory arrest, then invasive methods are needed. Extracorporeal rewarming is an invasive method, which can rapidly and safely rewarm core blood.³⁻⁵

Venovenous haemofiltration is readily available now in most district general hospitals where it can be used as an alternative to arteriovenous haemofiltration in treating patients with acute renal failure. It is an extracorporeal circulation that is an efficient rewarmers of core blood and thus, can be used to rewarm severely hypothermic patients who need more invasive measures.

Case history

A 77 year old woman found collapsed at home was admitted to hospital with a Glasgow Coma Scale (GCS) of 8 and rectal temperature of 26°C. Blood pressure was 93/55 and heart rate 89 in sinus rhythm. While rewarming with a heated blanket and warmed intravenous fluids she had a prolonged ventricular fibrillation (VF) arrest with episodes of other serious arrhythmias. Defibrillation was used while conventional rewarming methods continued, together with gastric lavage and bladder catheter irrigation, but with no success.

Even after five hours the core temperature was still only 30°C and she was continuing to have episodes of VF arrest that responded to defibrillation. As the cause of her obtunded conscious was not clear (GCS remained 7/15), and adrenaline (epinephrine) was maintaining her blood pressure, we intended to warm her efficiently to normothermia using venovenous haemofiltration in the A&E department. A temperature of 34°C was reached after one hour, her blood pressure was now 120/73 with adrenaline and her heart rate 111 in sinus rhythm.

As her GCS remained the same with mild hypothermia, she was referred to the medical team for further investigations towards a working diagnosis of a cerebral vascular accident, which caused her to collapse two days before admission. She died a day later.

Haemofiltration

Haemofiltration was performed using the Gambro AK-10 "Low flow" haemofiltration machine with BMM 10-1 Blood Monitor and HFM 10-1 Fluid Monitor. A vascath was inserted into the right femoral vein. The haemofiltration fluid was Normosol with 60 mmol of potassium added and the exchange volume was set to 27 litres with a filtration rate of 84 ml/min. Heparin was used as the anticoagulant (10 000 units in 40 ml normal saline run at 4 ml per hour). The Haemofilter was a polysulfone filter made by Link Medical/Bellco.

Because of the previous large warmed fluid transfusions, the machine was set to take off one litre of fluid to prevent overload. Haemofiltration was stopped after one hour having reached the desired temperature of 34°C.

Discussion

This case shows a practical and safe method of warming severely hypothermic patients using venovenous haemofiltration in the A&E department.

Venovenous haemofiltration is one form of extracorporeal rewarming. Successful outcomes with rewarming hypothermic patients have been reported with cardiopulmonary bypass,^{3,4} haemodialysis⁶ and peritoneal dialysis.⁷

For severe hypothermia with circulatory arrest, rewarming with cardiopulmonary bypass is the most efficient by being rapid and providing adequate and immediate circulatory support.^{4,5,8} However, it requires considerable time to set up (sometimes, exceeding one hour) and technical support. Its use is therefore often limited to operating theatres.

Continuous arteriovenous haemofiltration has also been successfully used to rewarm hypothermic patients.⁹ However, at our district general hospital, venovenous haemofiltration, involving insertion of a single large double lumen central venous catheter has been found to be more practical. A roller pump is used instead of the patient's blood pressure, to circulate the blood through the filtration unit before it is warmed and returned to the patient. Continuous venovenous rewarming has been successfully used in both a juvenile animal model,¹⁰ and in adult intensive care patients.⁵

Extracorporeal venovenous rewarming warms core blood directly and thus is more efficient than standard rewarming techniques (for example, warming blankets, warmed intravenous fluids, gastric, bladder and peritoneal lavage).⁵

Haemofiltration is now commonly used across the country in treating acute renal failure patients and in intensive care units. The haemofiltration machine is mains driven, easy to operate and very portable. It can be set up and ready to use in 20 minutes.

The experienced operator (intensivist/nephrologist/haemofiltration nurse) needs to set the fluid volume to be exchanged, the required fluid loss and the filtration rate (ml/min). The machine will then automatically carry out the treatment when activated, with the nurse supervising all data and checking the results. In our case, minimal filtration was required as the primary use was for rewarming. Treatment was terminated early as the desired temperature was reached.

Complications of haemofiltration are rare. For example air embolism, hypotension, infections, leaks in the filter or anticoagulation problems.

This case report shows the effectiveness and simplicity of using venovenous haemofiltration in the A&E setting.

- 1 Gentilello LM. Advances in the management of hypothermia. *Surg Clin North Am* 1995;75:243-56
- 2 Danzl DF, Pozos RS. Accidental hypothermia. *N Engl J Med* 1994;331:1756-60.
- 3 Dobson JAR, Burgess JJ. Resuscitation of severe hypothermia by extracorporeal rewarming in a child. *J Trauma* 1996;40:483-5.
- 4 Walpoth BH, Walpoth-Aslan BN, Mattle HP, et al. Outcome of survivors of accidental deep hypothermia and circulatory arrest treated with extracorporeal blood warming. *N Engl J Med* 1997;337:1500-5.
- 5 Gregory JS, Bergstein JM, Arahamian C, et al. Comparison of three methods of rewarming from hypothermia: Advantages of extracorporeal blood warming. *J Trauma* 1991;31:1247-52.
- 6 Hernandez E, Praga M, Alcazar JM, et al. Haemodialysis for treatment of accidental hypothermia. *Nephron* 1993;63:214-16.
- 7 Vella J, Farrell J, Leavey S, et al. The rapid reversal of profound hypothermia using peritoneal dialysis. *Ir J Med Sci* 1996;165:113-14.
- 8 Althaus U, Aeberhard P, Schupbach P, et al. Management of profound accidental hypothermia with cardiorespiratory arrest. *Ann Surg* 1982;195:492-5.
- 9 Broderson HP, Meurer T, Bolzenius K, et al. Haemofiltration in very severe hypothermia with favourable outcome. *Clin Nephrol* 1996;45:413-15.
- 10 Seigler RS, Golding E, Blackhurst DW. Continuous venovenous rewarming: results from a juvenile animal model. *Crit Care Med* 1998;26:2016-20.

Snap without crackle or pop: a rude awakening. A case history of penile fracture

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Abstract

Penile fracture is a rare but worrying condition. The presentation to accident and emergency or primary care should not present difficulty in diagnosis but may cause concern with regard to initial treatment and definitive management. Emergency admission to a urologist is mandatory.

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Keywords: penile fracture; urology

Case report

A 32 year old white man presented to the accident and emergency (A&E) department of Bradford Royal Infirmary complaining of discomfort and bruising of his penis over a four hour period. That morning he had awoken with an erection, he wanted to pass urine and as a result had forcefully bent his penis. On bending the shaft of his penis he heard a “loud snap” and he experienced pain, loss of penile tumescence and swelling and bruising of his penis. His partner who was in the next room also heard the “snap” and insisted that they go to an A&E department. The swelling of the penis increased up until the time of assessment.

Examination in the A&E department revealed a flaccid penis with a large fluctuant haematoma on the dorsal aspect of his penis stretching from the base of the shaft to the glans. There was some tenderness on palpation. The patient had micturated successfully since sustaining the injury without any frank haematuria.

Investigations performed included a full blood count, coagulation profile and urea and electrolytes, all of which were normal.

The patient was referred urgently to the urology service.

Discussion

Fracture of the penis has attracted interest in the medical literature from many countries including Zimbabwe, Taiwan, Scandinavia, India, Iran, Hungary, the United States and England.¹ Penile fracture is caused by a traumatic rupture of the tunica albuginea of one or both corpora cavernosa. The fracture is most frequently reported as happening to the erect penis during sexual intercourse, but it has been reported after manipulation and falling onto the erect penis.

In St George's Hospital in London 0.25% of all emergency urology admissions are attributable to penile fracture.² The differential diagnosis includes a tear of the deep dorsal vein of the penis. Classically the condition presents

with a sudden snap, pain, detumescence and a haematoma of the penis with deformity. The “snap” heard when the tunica albuginea tears has been well described, as has the fact that it is also often heard by the sexual partner if the fracture occurs during coitus. To our knowledge this is the first time that the snap has been reported as having been heard by a partner not in the same room at the time of fracture. Clinical examination consistently reveals a smooth, fixed, tender, palpable lump at the fracture site. This lump apparently is formed by a clot in the torn corpus cavernosum, trapped in its well localised position by Buck's fascia.³ Investigations suggested for the condition include ultrasonography, cavernosography, magnetic resonance imaging, urethrography and surgical exploration. Dissection in this condition tends to reveal a tear in the proximal third of the corpus cavernosum.¹

Complications of the disorder include concomitant urethral injuries (20%), penile curvature, abscess formation, penile pain, pulsatile diverticulum, urine extravasation, psychophysical problems, and impotence.^{3,4} Urethral injury should be suspected if there is blood at the meatus or failure to pass urine.

Treatment may be conservative or operative. Conservative regimens include catheterisation once a urethral injury has been ruled out, pressure dressings, penile splinting, diazepam, non-steroidal anti-inflammatory drugs and even oestrogen therapy.⁵ The rationale for the use of diazepam is that it reduces the frequency and intensity of erections and so lessens discomfort.

The more recent literature seems to favour operative management to lessen the likelihood of complications that are more common in the conservatively treated group, up to 53% in the study by Kalash *et al.*⁶ The operative management of the condition ideally is as soon as possible.⁷ The surgical approach may include a procedure under local anaesthesia or the more extensive circumferential or longitudinal incision and degloving of the penis to evacuate the haematoma and locate and repair the tear.^{3,4}

Conclusion

Penile fracture is a rare condition but it merits sensitive handling and urgent referral to a urologist for further management.

Funding: none.

Conflicts of interest: none.

¹ Asgari MA, Hosseini SY, Safarinejad MR, *et al.* Penile fractures: evaluation, therapeutic approaches and long-term results. *J Urol* 1996;155:148–9.

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- 2 Morris SB, Miller M, Anson K. Management of penile fracture. *J R Soc Med* 1998;91:427–8.
- 3 Naraysingh V, Maharaj D, Kuruvilla T, et al. Urology. Simple repair of fractured penis. *J R Coll Surg Edinb* 1998;43:97–8.
- 4 Nicoliasen GS, Melamud A, McAnnich JW. Rupture of the corpus cavernosum: surgical management. *J Urol* 1983;130:917–19.
- 5 Abulata KA, Awad RA. Fracture shaft of penis: Non surgical treatment of three cases. *J R Coll Surg Edinb* 1983;28:266–7.
- 6 Kalash SS, Young TD. Fracture of the penis: controversy of surgical versus conservative treatment. *Urology* 1984;24:21–4.
- 7 Cummings JM, Parra RO, Boulter JA. Delayed Repair of penile fracture. *J Trauma* 1998;45:153–4.

Brugada syndrome—the missed epidemic

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Abstract

About 10–20% of patients dying suddenly or resuscitated from ventricular fibrillation do not have demonstrable heart disease. These people are often young and tragically in some cases sudden death is the first and only clinical event. One of the three main electrophysiological diagnoses to be considered in these situations is the Brugada syndrome. A case of Brugada syndrome is described, together with an example of the classic electrocardiographic manifestations and a discussion of the possible aetiology, diagnosis and management of this condition.

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Keywords: Brugada syndrome

Case report

At approximately 6 pm one evening, a 29 year old man presented at the accident and emergency (A&E) department in a critical condition. The history obtained from both the paramedics and police was unusual. The policeman had been driving his car on the motorway when the car in front, carrying a single occupant, suddenly veered off the road, crashed through a barrier and came to a stop in a field. The policeman found the driver to be in cardiorespiratory arrest, and started resuscitation at the scene. On arrival, the paramedics discovered the driver's initial cardiac rhythm to be ventricular fibrillation and he was success-

fully defibrillated. During transfer to the A&E department he required defibrillation for a further episode of ventricular fibrillation.

On arrival in hospital he was agitated, with a Glasgow Coma Score of 8/15 but was breathing spontaneously with a partially obstructed airway. The monitor showed normal sinus rhythm with an initial blood pressure of 130/80. He was given a rapid sequence induction of anaesthesia to maintain and protect his airway. Initial primary and secondary trauma surveys revealed no obvious injury and initial chest, pelvis, and c-spine radiographs were unremarkable. Initial speculative diagnoses included spontaneous intracerebral haemorrhage, cardiac arrhythmias and convulsion. The patient had urgent computed tomography of the head, which was normal. Further history from the family revealed no previous medical problems and no relevant drug history.

The initial 12 lead electrocardiograph (ECG) (see fig 1) showed a partial right bundle branch block with ST segment elevation in chest leads V1 to V3. At this stage a primary cardiac problem was thought to be more likely. Urgent echocardiography to look for structural cardiac disease was performed. No abnormality was found. Throughout this time period the patient maintained a normal sinus cardiac rhythm. He was transferred to an intensive care bed and weaned uneventfully from ventilation over a period of 24 hours. Toxicology screening was negative.

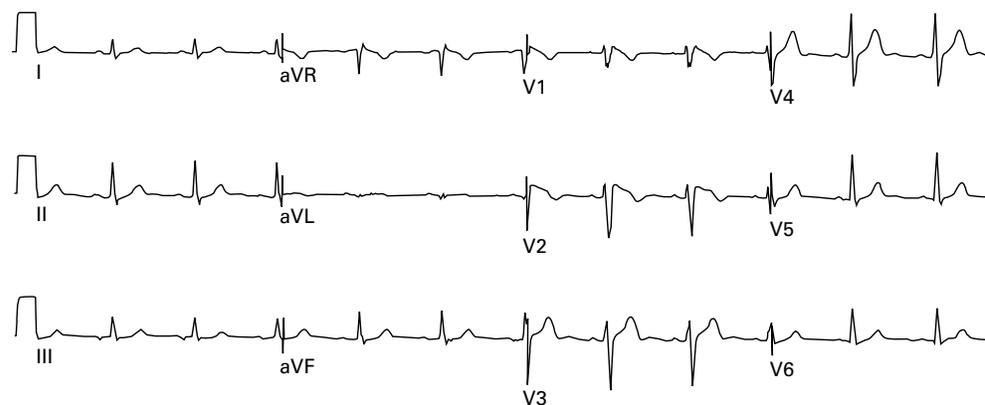


Figure 1 ECG showing classic ST segment elevation in chest leads V1–V3 with right conduction delay.

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After extubation he was neurologically intact and was transferred to the care of the cardiologists for further investigation. Coronary angiography was carried out to exclude coronary artery disease and congenital artery anomalies. None were found. Right ventricular angiography showed no impairment of function. The clinical and electrocardiographic features confirmed the diagnosis of Brugada syndrome. An implantable defibrillator was inserted within days of presentation and the patient was discharged home.

Subsequent follow up has been unremarkable with no further episodes of arrhythmias.

Discussion

About 10–20% of patients dying suddenly or resuscitated from ventricular fibrillation do not have demonstrable heart disease.¹ Unexpected arrhythmogenic death occurring in people with minimal or no structural heart disease is estimated to represent 3% to 9% of out of hospital cases of ventricular fibrillation unrelated to myocardial infarction.² These patients are often young and tragically sudden death may be the first and only clinical event.

When acute ischaemia is not the cause of sudden cardiac arrest, there are three main electrical cardiac disorders to be considered as the possible cause of ventricular fibrillation: the Brugada syndrome, the long QT syndrome, and the Wolff-Parkinson White syndrome. If these three disorders are excluded, and the heart is structurally normal, then ventricular fibrillation is considered idiopathic.

In 1992 Pedro and Josep Brugada³ described eight otherwise healthy patients with sudden and aborted cardiac death, in whom they found “right bundle branch block and persistent ST segment elevation in leads V1 to V3”. Based on these observations they outlined a new distinct clinical and electrocardiographic syndrome. This was subsequently called the Brugada syndrome. In these patients the ECG showed right bundle branch block, normal QT

interval and persistent ST elevation in the precordial leads V1 to V2–V3 not explainable by electrolyte disturbances, ischaemia or structural heart disease. All these patients had right bundle branch block. Repolarisation was abnormal, characterised by persistent ST elevation (at least 0.1 mV) in leads V1 to V2–V3. The PR and QT intervals were within normal limits. ECG features of right bundle branch block with ST elevation in the right precordial leads has subsequently been described as the “Brugada sign”.⁴

ST segment elevation in the right chest leads is observed in a variety of clinical and experimental settings and is not unique or highly specific for the Brugada syndrome. A clear distinction of this syndrome cannot be made on the basis of the ECG alone. The prevalence of idiopathic ST segment elevation is reported to be 2.1% to 2.65%. An elevated ST segment limited to the right precordial leads occurs in less than 1% of all cases of elevated ST segment.² However, ST segment elevation in the right precordial leads in the absence of ischaemia, electrolyte or metabolic disorders, pulmonary or inflammatory disorders or abnormalities of the central or peripheral nervous system is suggestive of the Brugada syndrome. In many cases of Brugada syndrome, the ECG manifestations can normalise transiently, leading to underdiagnosis of the syndrome.² Strong sodium channel blocking agents such as procainamide and flecainide can unmask the ST segment elevation in many patients thus aiding diagnosis.

Brugada reported that malignant arrhythmias eventually occur in 27% of initially asymptomatic patients who have the Brugada sign. On the basis of reports published it has been speculated that 40% to 60% of patients diagnosed as having idiopathic ventricular fibrillation may actually suffer from Brugada syndrome.⁵ It is now thought to be the most frequent cause of sudden cardiac death in

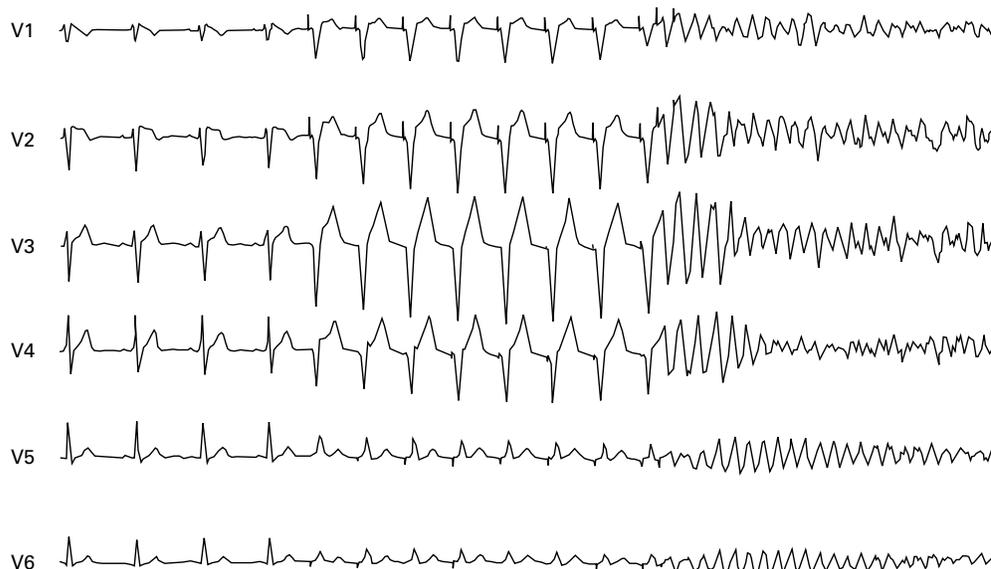


Figure 2 ECG of patient with Brugada syndrome showing classic precordial ST elevation followed by the development of ventricular fibrillation.

KEY POINTS

- Brugada syndrome is a cause of sudden cardiac death in previously healthy young people.
- ECG characteristics include: ST elevation in pre-cordial leads (V1–V3), right ventricular conduction delay with a normal QT interval.
- These patients require early cardiology opinion and electrophysiological investigation.
- Treatment consists of implantable defibrillators to prevent sudden death from ventricular fibrillation.

patients without structural heart disease under the age of 50 years.

The mean age of affected people is mid to late thirties. In the majority of cases tachyarrhythmias occur at rest and in many cases during the night. The recurrence rate of new arrhythmic events is as high as 40% in these patients.⁵

Current available data suggest that the Brugada syndrome is a primary electrical disease resulting in abnormal electrophysiological activity in the right ventricular epicardium. Electrical heterogeneity within the right ventricular epicardium leads to the development of coupled premature ventricular contractions via a re-entrant mechanism that precipitates ventricular tachycardia/fibrillation (see fig 2). The role of right bundle branch block in Brugada syndrome has been a matter of controversy. Conduction delay in the right ventricle does not seem to be an integral part of the syndrome. Furthermore, there seems to be no correlation between right bundle branch block and cardiac death, whereas a definite link exists between the magnitude of ST elevation and the incidence of the life threatening arrhythmias seen in these patients.²

The aetiology of Brugada syndrome is controversial. Brugada and Brugada suggested no structural abnormality of the heart, claiming the syndrome was attributable to the presence of a functional electrical disease.⁶ Martini *et al* suggested the presence of concealed right ventricular myocardial disease. Corrado *et al* report that a familial cardiomyopathy that mainly involves the right ventricle and conducting syndrome is responsible.⁷ An ion channel defect resulting in heterogeneous loss of the action potential dome in right ventricu-

lar epicardium has been proposed as a mechanism for Brugada syndrome. Three different mutations of the gene of the sodium channel SCN5A on chromosome three have been identified.⁸ The Brugada syndrome is autosomal dominantly inherited but has variable expression. Testing of all family members is important because of the high incidence of familial occurrence.

Patients with Brugada syndrome have a high incidence of sudden death, and prophylactic defibrillators are indicated in those with inducible arrhythmias at electrophysiological study, irrespective of symptoms.⁹ In contrast, the incidence of sudden death in the long QT syndrome is very low, making prophylactic defibrillator implantation not cost effective. Even with the best medical treatment, arrhythmia recurrence rates are still 45–50% at five years. Pharmacological treatment does not protect against recurrent events and implantation of a cardiac defibrillators is the only proven effective treatment in preventing sudden death in patients with the Brugada syndrome.^{10 11} Emergency physicians therefore have an important role in referring suspected cases of Brugada syndrome for urgent cardiology opinion for consideration of an implantable defibrillator.

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- 1 Zipes D, Wellens HJJ. Sudden cardiac death. *Circulation* 1998;**98**:2334–51.
- 2 Gussak I, Antzelevitch C, Bjerregaard P, *et al*. The Brugada syndrome. clinical, electrophysiologic and genetic aspects. *J Am Coll Cardiol* 1999;**33**:5–15.
- 3 Brugada P, Brugada J. Right bundle branch block, persistent ST segment elevation and sudden cardiac death: a distinct clinical and electrocardiographic syndrome. *J Am Coll Cardiol* 1992;**20**:1391–6.
- 4 Viskin S, Belhassen B. Polymorphic ventricular tachyarrhythmias in the absence of organic heart disease: Classification, differential diagnosis, and implications for therapy. *Prog Cardiovasc Dis* 1998;**41**:17–34.
- 5 Alings M, Wilde A. “Brugada” syndrome. Clinical data and suggested pathophysiological mechanism. *Circulation* 1999;**99**:666–73.
- 6 Brugada P, Brugada J. Right bundle branch block and ST segment elevation in leads V1–V3. A marker for sudden death in patients with no demonstrable structural heart disease. *Circulation* 1998;**97**:457–60.
- 7 Tada H, Aihara N, Ohe T, *et al*. Arrhythmogenic right ventricular cardiomyopathy underlies syndrome of right bundle branch block, ST segment elevation and sudden death. *Am J Cardiol* 1998;**81**:519–22.
- 8 Kirsch GE, Zhang D, Brugada R, *et al*. Genetic basis and molecular mechanisms for idiopathic ventricular fibrillation. *Nature* 1997;**392**:293–6.
- 9 Brugada P, Brugada R, Brugada J, *et al*. Use of the prophylactic implantable cardioverter defibrillator for patients with normal hearts. *Am J Cardiol* 1999;**83**:98–100D.
- 10 Causser J, Connelly D. Implantable defibrillators for life threatening ventricular arrhythmias. *BMJ* 1998;**317**:762–3.
- 11 The AVID Investigators. A comparison of anti-arrhythmic drug therapy with implantable defibrillators in patients resuscitated from near-fatal ventricular arrhythmias. *N Engl J Med* 1997;**337**:1576–83.