

Hypertension secondary to Marfan's syndrome initially attributed to an anxiety disorder

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CASE REPORT

A 15-year-old boy was admitted to our inpatient unit for assessment and treatment after a 2-year history of restricted exercise tolerance, tiredness, impaired concentration, forgetfulness, myalgia and sleep disturbance following bacille Calmette-Guérin vaccination. He had previously attracted a label of chronic fatigue syndrome.

Mental state examination revealed subjective feelings of anxiety, muscle aches, difficulty getting off to sleep, nocturnal waking, low mood, frustration, apprehension relating to every day events and inability to relax. Physical concomitants of anxiety were also marked including trembling, restlessness, nausea and vomiting.

Physical examination revealed pectus excavatum, high arched palate, joint hypermobility and arachnodactyly. He was noted to be tall 1.82m (95th centile) and thin, weight 50.9 kg (between 25th and 50th centile).

In view of the Marfanoid features an ophthalmology opinion was sought, which revealed no abnormality. A hand X-ray was taken (*Figure 1*) which demonstrated a metacarpal index of 9 which is consistent with Marfan's syndrome. An echocardiogram previously performed at another unit was reported to be normal.

A clinical geneticist then reviewed the patient, but felt that a diagnosis of Marfan's syndrome could not be made based on his skeletal features alone. No cardiac murmur was audible at this time. Blood pressure (BP) on admission was 139/80 mmHg (Dynamat, Johnson and Johnson). This was attributed at the time to the stress of hospital admission.

While on the ward it was ascertained that he became extremely anxious in social situations, especially school assembly and other events relating to large crowds. This was initially treated with a behavioural programme which included anxiety management, social skills and assertiveness training. However, when reintegration to his own school was proposed his symptoms became exacerbated. He was increasingly anxious, had difficulty getting off to sleep, had 'panic attacks', developed poor appetite alongside intermittent vomiting, had diminished food intake and lost weight.

A tricyclic antidepressant, trimipramine, gradually increased to 75 mg a day, was added to the treatment regimen because of significant low mood. There was some improvement, but the anxiety symptoms persisted. It was therefore elected to recheck his BP before considering a β -blocker.

All subsequent readings over the next 2 days were high: 140/80 mmHg, 150/90 mmHg, 145/90 mmHg, 155/90 mmHg, 150/90 mmHg, 140/100 mmHg (all measurements taken manually by one person).

Further investigations were thus performed. Electrolytes, urine amino acids, thyroid function tests, abdominal ultrasound and electrocardiogram were all normal. Chest X-ray was unremarkable and karyotype was 46 (X,Y).

Medication was commenced initially with propranolol, but his hypertension became more pronounced (170/90 mmHg, 160/95 mmHg, 165/100 mmHg) so this was changed to labetalol with therapeutic benefit. (24-hour urine catecholamines were later shown to be normal.)

In view of our continued concern about his raised BP and physiognomy, it was elected to refer him to our cardiology team. The echocardiogram was repeated which demonstrated mild mitral valve prolapse alongside mild aortic root dilatation of 3.3 cm against a left atrial dimension of 1.77 cm.

Medication was therefore changed to atenolol 25 mg twice daily. Subsequently, his hypertension settled and his anxiety state became less pronounced. He achieved reasonable GCSE results and went on to sixth form college. He remains well without any further psychiatric intervention.

INTRODUCTION

Among the multitude of associated symptoms of secondary high blood pressure, anxiety is common. However, underlying organic causes should always be sought in children before repeated high blood pressure measurements are dismissed as merely reflecting an anxiety disorder. Anxiety symptoms themselves could have a physical basis and mitral valve prolapse has been implicated as one such possibility.

DISCUSSION

By having both cardiovascular and musculoskeletal features our patient fulfils the diagnostic criteria for Marfan's syndrome (Lipscomb et al,



Figure 1. X-ray of patient's hand.

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1997). Marfan's syndrome is a known cause of both mitral valve prolapse and aortic root dilatation, reflecting the underlying defective production of fibrillin on chromosome 15 (Kainulainen et al, 1990; Dietz et al, 1991).

Our patient also fulfils the criteria for a generalized anxiety disorder with additional features of a phobic anxiety state (*International Classification of Disorders 10*, World Health Organization, 1993). His age and the raised blood pressure recordings over several weeks are indicative of significant hypertension (Report of the second task force on blood pressure control in children, 1987). Trimipramine can raise the diastolic blood pressure but the consistently high blood pressure recordings, both systolic and diastolic, in our patient cannot be totally attributed to this medication.

Raised blood pressure can be one feature of an anxiety state or can reflect underlying organic pathology. Aortic root dilatation can be a consequence of hypertension. However, in our patient features of anxiety and depression can also be secondary to mitral valve prolapse; anxiety, panic and depressive symptoms being well

recognized aspects of this condition (Vitiello et al, 1990; Stavrakaki et al, 1991). In this case, treatment of hypertension with β -blockers also had a secondary benefit in terms of the physical concomitants of anxiety, thus augmenting the psychological treatment.

Our patient also has hypermobility of the joints. This finding could also have some link with the anxiety symptoms (Bulbena et al, 1993). The association between anxiety disorders and joint hypermobility has been recognized in adults. However, there does not appear to be any literature that relates to children.

CONCLUSIONS

A recent review of Marfan's syndrome in children (Lipscomb et al, 1997) did not describe any psychiatric elements of this condition. We were unable to find any relevant literature linking anxiety symptoms and Marfan's syndrome in any way other than mitral valve prolapse. Organic illness often manifests as apparent psychiatric symptomatology as with our patient. Therefore, psychiatric and medical teams need to work closely together to care for their patients appropriately and holistically.

Although anxiety symptoms are extremely common in children due care and attention should be given to the possibility of any underlying organic pathology. Arriving at the correct underlying diagnosis and hence appropriate management can inspire patient confidence and compliance. Paradoxically, having a diagnosis (even one carrying impaired life expectancy) can relieve anxiety. **HM**

Bulbena A, Duro' JC, Porta M et al (1993) Anxiety disorders in the joint hypermobility syndrome. *Psychiatry Res* **46**: 59-68

Dietz HC, Cutting GR, Pyritz RE (1991) Marfan's syndrome caused by a recurrent de novo missense mutation in the Fibrillin gene. *Nature* **352**: 337-9

Kainulainen K, Pulkkinen L, Savolainen A, Kaitila I, Peltonen L (1990) Location of chromosome 15 of the gene causing Marfan's syndrome. *N Engl J Med* **323**: 935-9

Lipscomb K, Clayton-Smith J, Harris R (1997) Evolving phenotype of Marfan's syndrome. *Arch Dis Child* **76**: 41-6

Report of the second task force on blood pressure control in children (1987) The National Heart, Lung and Blood Institute, Bethesda Maryland. *Paediatrics* **79**(1): 1-25

Stavrakaki C, Williams E, Boisjoli A, Vlad P, Chasse H (1991) Mitral valve prolapse and psychiatric complications—a case report. *J Psychiatry Neurosci* **16**(1): 45-9

World Health Organization (1993) *The ICD-10 Classification of Mental and Behavioural Disorders—Diagnostic Criteria for Research*. World Health Organization, Geneva

Vitiello B, Behar B, Wolfson S, McLeer SV (1990) Diagnosis of panic disorder in prepubertal children. *J Am Acad Child Adolescent Psychiatry* **29**(5): 782-4

IN THE PUBLIC'S VIEW...

Putting the brakes on child traffic casualties

It is the age of accountability and responsibility. We cannot let practices continue that harm people: we must take action, we must not be afraid to question and challenge our colleagues, and report them, if necessary, for sub-standard practice.

So next time you are in your colleague's car and they are exceeding the speed limit, just remind them of the limit and ask them to slow down. Or try telling your next taxi driver. When they ignore you, tell the police.

Panorama (BBC1, 14 February) told the stark truth: children are killed because cars are safe for their drivers. Hit a child at 20 mph and one in twenty dies; at 40 mph 17 in 20 die. It's not company car drivers 'doing a ton' in the outside lane of motorways that kill children; it's you and I nipping between hospitals at 40 mph in a 30 mph limit. Check your speedo next time you're late for a clinic.

Panorama spent too much time talking to parents who'd lost their children and not enough

defining the problem. No one boasts or jokes any more about driving home after downing 5 pints and half a bottle of whisky. Strictly teetotal drivers are no longer considered wimps. But try telling a group of colleagues that you never (or hardly ever) break speed limits. If you are believed, you are likely to be thought a goodie-goodie. There was much sympathy for a colleague prosecuted for driving at 55 mph in a 40 mph zone just near the hospital — mainly on the lines of: 'Haven't the police got anything better to do?' A little while later I came across the result of an accident on that same stretch of road; the last time I enquired the 8-year-old was still unconscious on the neuro-ICU.

So I freely admit, I'm (almost) a goodie-goodie. I don't drive more than a few miles per hour more than the speed limit, and in town there is a constant stream of traffic overtaking me. I make no claim that I am a good driver, but at least if I am unlucky enough or even foolish enough to hit a child, I am

perhaps less likely to do them serious harm. I don't tap taxi drivers on the shoulder, however.

This government has apparently declared somewhere between 600 and 800 targets for the public services since coming into office. As with targets in the NHS, in other services targets are all we get; the means to achieve the targets are left to the imaginations of the public servants. I've no doubt that one government target is to cut child casualties on the roads, and there is one clear way the government can show it means business at no financial cost. In all my driving around in Bristol, I have never once encountered a kangaroo, or for that matter a bull — so why haven't bull-bars on off-road vehicles been banned? If the government carried through this simple, sensible, effective measure, we might even listen more carefully to their ideas for the NHS. **HM**

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