Reviewer's report

Title: Postural orthostatic tachycardia syndrome after surgical correction of an aortic coarctation: a case report.

Version: 1 Date: 24 October 2011

Reviewer: Jonathan Johnson

Which of the following best describes what type of case report this is?: An unexpected association between diseases or symptoms

Has the case been reported coherently?: Yes

Is the case report authentic?: Yes

Is the case report ethical?: Yes

Is there any missing information that you think must be added before publication?: Yes

Is this case worth reporting?: Yes

Is the case report persuasive?: No

Does the case report have explanatory value?: No

Does the case report have diagnostic value?: Yes

Will the case report make a difference to clinical practice?: Yes

Is the anonymity of the patient protected?: Yes

Comments to authors:

Fernex et al present an interesting case report of a 29 year old male who was diagnosed with both postural orthostatic tachycardia syndrome (POTS) and upper airway resistance syndrome (UARS) 18 months after aortic coarctation repair. They assert that this is a causal relationship, and also report a novel finding of POTS and UARS in the same patient.

At best, this case report is a clear overstating of the case. There are many details that are left out that are critical to the case, while the discussion that is provided does not adequately theorize the mechanism behind this patient’s presentation. The case is however interesting, and warrants publication if many major corrections are made.
Major suggestions:

1. I would suggest the authors remove the use of the term “causal”. Quite simply, there is no proof to this. I believe that they can report that this patient was diagnosed with POTS and UARS, with symptoms starting after surgical correction of a coarctation, but that it is far as one can take it.

2. POTS is known to occur in some patients after a “stressor” of some kind that causes them to be supine for a prolonged period of time. I would like to know how long after his surgery the symptoms started – did he ever fully recover from surgery? Was he able to work after surgery but then got worse?

3. What about the coarctation repair made this happen? The authors need to supply many more details about the type of coarctation, location of arch resection or manipulation, details of the surgery and surgical recovery, severity of the ascending aorta dilation (a known concern in patients with bicuspid aortic valves and coarctation), gradient before and after surgery, why the tube graft was chosen instead of an end-end repair or stent, BP concerns before surgery, medical treatment for hypertension before surgery, etc. When was the diagnosis made of coarctation? He had surgery at the age of 11 – had anyone examined his heart and pulses prior to this surgery? How bad were his thoracic collaterals prior to surgical repair due to the coarctation? What was his exercise tolerance before surgery? Were there any complications of surgery? What was his BP after surgery – he was being treated with multiple anti-hypertensives, and so I assume he had some residual hypertension.

4. The relationship with POTS and UARS, while the authors assert that it has not been reported, may be only a function of the relative novelty of both diagnoses. The fact that POTS and CFS have such an overlapping presentation, and that UARS occurs in patients with IBS and CFS, it was only a matter of time before some patient was given the diagnosis of both POTS and UARS. I suspect that if all POTS patients had sleep studies, we would find many more with UARS or other similar sleeping disorders. I suggest that the authors comment on the overlapping presentations of patients who ultimately are diagnosed with CFS, fibromyalgia, UARS, and POTS.

5. How much role did the micrognathia play in the UARS and sleep symptoms? The jaw abnormality was mentioned in the diagnosis but not discussed further in the paper.

6. Why were the results of the physical exam and the HUTT so different? In the exam room, his HR rose 70 bpm on standing for 5 minutes, but in the HUTT he barely made it to 30 bpm.

7. Why was the Holter performed? And what did the authors make of the repeated atrial tachycardia runs – were these considered significant? Were all 59 atrial tachycardia episodes 3 or 4 beats in length?

8. I am intrigued as to the rapid recovery. I don’t dispute the diagnosis of POTS, but typically our patients take much longer to recover. Isn’t it possible that simply removing the amlodipine and valsartan could have resolved the symptoms, since these are both known to exacerbate symptoms in patients with POTS or...
orthostatic intolerance? One of the primary treatments after POTS diagnosis is the removal of “offending agents”, which may have been the main therapy in him, especially considering how rapidly he improved.

9. The timeline of the presentation is difficult to follow. I would suggest the authors re-write the case report following the case from the beginning (before coarctation repair) to the end (recovery), following a straight timeline.

10. I am not sure there needs to be such an exhaustive description of POTS and UARS in the discussion – the authors could describe these entities in brief and use the extra space to discuss mechanisms behind how this patient could develop POTS after coarctation repair (there currently are only two lines about blood pressure issues after coarctation repair, nothing about heart rate or autonomic issues - ? due to time spent supine vs. deconditioning vs. meds, etc…….). No need to re-hash the 3 different subtypes of POTS which has been published extensively.

11. Table 1 is unnecessary – everything aside from the HR is normal.

12. What is the timeframe of the lithotripsy to the coarctation repair and the POTS presentation? Unclear if this was done prior to the coarctation repair or after. IF after, did this play a role in the presentation?

13. Did his symptoms change once he removed the cola-drink from his diet? Was this a caffeinated drink? Caffeine is known to exacerbate symptoms of POTS.

14. Is it possible that the UARS was present prior to surgery? Did his girlfriend note any difference in symptoms before and after coarctation surgery (i.e. did the sleep disturbance only start after surgery)?

Minor suggestions

Page 5, line 8 – “the patient could not stand for some minutes” – how many exactly?

Page 6, line 21 – “the presence of a POTS” – remove the word “a”

Page 8, line 1 – suggest not saying “misdiagnosed” with chronic anxiety or panic disorder, as both can be “co-diagnosed” with POTS.

Page 8, line 12-14 – this sentence is confusing – exactly what are the distinguishing features that the authors want to list – what do the BP, stroke volume, and HR due to tilt, what does the plasma norepi level do with tilt, etc.

Page 8, line 21: “10-15% of POTS experience” – suggest inserting the word “patients” before “POTS”.

Conclusion: When the authors say “this case report could help better understand its pathophysiology……”, this is not true of the manuscript in its current form – the authors need to expound more in the discussion on why they think this patient presented the way he did. Nothing new has been presented in terms of understanding the pathophysiology of POTS. Suggest removing this line.

Quality of written English: Needs some language corrections before being published
Declaration of competing interests:

I declare that I have no competing interests.