Author's response to reviews

Title: Adult diagnosis of Swyer-James-MacLeod syndrome: a case report.

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Version: 2 Date: 12 May 2010

Author's response to reviews: see over
[Cover Letter]

MS: 4486840293369673

Adult diagnosis of Swyer-James-Macleod syndrome: a case report.

Carlos Capela, Paulo Gouveia, Marco Sousa and Maria J Regadas

(A) Point-by-point response to Dr Manelaos Karanikolas

1. Case presentation: I think it is more appropriate to use past tense, rather than present tense (a patient “presented”, rather than “presents”, etc).

Response: Many corrections were made. All text was revised by a person truly proficient in American English.

2. Case presentation: can the authors be more specific about phlebotomy? I am assuming they removed blood from the patient. If so, how much? What was the indication? Why on two occasions? What was the end-point of this treatment?

Response: We made a 500 ml phlebotomy by the time we considered the present patient to have a polycythemia secondary to a cyanotic congenital heart disease mostly associated to the newly diagnosed patent ductus arteriosus with right-to-left shunt. The authors considered, that moment, a beneficial option to relieve symptoms. The second phlebotomy was performed later, in a situation of patient’s refusing in further care. The authors consider in present article revision, to omit this information.

3. The authors state that the patient finally had “cardiothoracic consultation”. Was that cardiothoracic surgery consultation, or what? What was the conclusion and recommendations from this consultation?

Response: A correction was made. We were talking about a Cardiothoracic Surgery Consultation. The patient ultimately denied further evaluation, including that consultation. It’s referred later on the text and the authors considered no to repeat this point.

4. I think the Discussion is too short, whereas the Conclusion is too long. I think most of what the authors have included in “conclusion” should really be included in “discussion”.

Response: A correction was made.

5. In my opinion, this report deserves publication. However, can the authors explicitly, clearly state what is unique or important about this case report, and why they think this case is worth publishing?
Response: A correction was made. The authors included new relevant information contributing to emphasize the why this case is worth publishing.

6. I think the 2nd paragraph of “Conclusion” belongs to “Discussion”. Furthermore, I also think this paragraph needs to be expanded, to include a more thorough discussion of what is known about this rare syndrome. As it is now, after reading this paragraph, I still do not know how rare the syndrome is. Are there a few reports, or a few hundred reports worldwide? How about incidence or prevalence data for the entire population? Does it occur worldwide, or just in Europe, or just, perhaps, in industrialized nations? Natural history of the disease? Morbidity and/or mortality data?

Response: Fewer than 5 cases were reported in last 10 years (www.pubmed.com) and no epidemiological science was possible with those numbers; about morbidity and mortality associated to SJMS was related to higher risk of infection and chronic lung disease, characterized by bronchiolar abnormality but no major morbidity-mortality studies were made with SJMS.

7. The text has several minor errors, mostly related to expression. For example, the authors state “his” physical examination, but they are discussing a female patient. In addition, some sentences need simplification and/or improvement. The authors also use “a” even in places where it is unnecessary or wrong (for example: “It is characterized by an hypoplasia and/or agenesis....” It is better to simply say: “It is characterized by hypoplasia and/or agenesis....”). As this is only one of many similar disruptive (but not fatal) errors in this document, I would recommend that the authors have a person truly proficient in American English read the entire manuscript, in an attempt to improve clarity, simplify some expressions and eliminate most of these minor errors.

Response: Corrections made. The all text was revised by a person truly proficient in American English.

8. The figures are meaningful and enhance the text. Could the authors provide a meaningful legend for figure 1?

Response: Some corrections were made. The first interpretation of pulmonary x-ray wasn’t the most correct as revealed with complemented radiological studies and this is one of the main characteristics of such disease; authors consider no to reveal, at this point, a meaningful to the figure 1.

9. Legends in figures 2 and 3 state the type of test twice. Eliminating “Chest HRCT. Legend” from the legend in figure 2, and eliminating “Chest CT angiography. Legend:” in figure 3 would simplify these legends while presenting exactly the same meaningful information.

Response: Some corrections were made.
(B) Point-by-point response to Doctor Indranil Chakravorty

1. I would recommend adding more quantitative data on echocardiographic function including dimensions of the right sided chambers, estimated pulmonary arterial pressure and an electrocardiogram showing the right sided ‘strain’ pattern described.

Response: The authors agreed about the relevance of adding more quantitative data and some corrections were made. Considering the number and utility of pictures that should accompany the text, the authors considered not include the electrocardiogram.

2. The formal laboratory lung function test demonstrating a mixed obstructive and restrictive picture needs to be shown and the underlying pathophysiology explained to the reader

Response: The well described Chest High-resolution Computed Tomography Scan revealing hyperlucency and diminished vascularity in the right lower and middle lobe with hyperinflation of the pulmonary parenchyma as well as a diminished right pulmonary vascular beds which was confirmed by the CT angiography (Angio-CT) by a diffuse hypoplasia / agenesis of right superior and inferior branches of the pulmonary artery could perfectly explain the obstructive-restrictive pattern seen on pulmonary test function.

3. The shunt through the PDA must have contributed to the development of the pulmonary arterial hypertension and the presence of a murmur and the Doppler demonstration of continuous flow is significant. This shunt can be estimated by the reduction in gas transfer from formal laboratory lung function. The lack of normalization in arterial hypoxia in the presence of oxygen supplementation may also be a non-invasive confirmation of the contribution of the PDA to the development of PAH (as the patient refused cardiac catheterisation studies).

Response: Relevant data was added to the text demonstrating that.

4. Trans-oesophageal echocardiography may be able to provide better quality data as the ‘normal heart chamber dimensions’ described by the authors is unlikely to be compatible with the severity of clinical signs. I would recommend that this is obtained, if feasible.

Response: Some corrections were made concerning this point. The patient also refused trans-oesophageal echocardiography study by the moment we determine some degree of PAH with no repercussion in right side heart chambers dimensions seen on TTE.

5. In the end, the authors acknowledge that the 3 factors; which may have contributed to the patients’ presentation (Obstructive lung disease due to obliterative bronchiolitis, differential pulmonary blood flow due to SJM and shunt through a PDA) would be difficult to quantify.

Response: Authors demonstrate that concern and pointed that a cardiac catheterization would be necessary the better clarify the relative contribution of each factor.

6. Therefore, I would recommend acknowledging this in the title and the abstract of this article to reflect ‘SJM presenting late with right heart failure secondary to pulmonary arterial
hypertension complicated by a PDA’. This would then reflect the entire complexity of this case and the discussion will need to recognize and explore this interaction further.

Response: This was probably the most difficult point we had to response. The authors ultimately consider not add complexity into the title or in the abstract as our mainly objective was to report a rare medical condition with one unusual associated aspect – being recognized 10 to 15 years later; This is the main scope of the journal. Anyway, some changes were made and adding a second unusual aspect, (PDA diagnosis) wouldn’t turn it, at a first glance, more complex.

7. The discussion may need to acknowledge another case report of VSD and SJM co-presenting with reversal of shunt (Int J Cardiol. 2006 Oct 26;113(1):E4-6) and discuss the contribution of the possible shunt in such cases.

Response: The authors knew already this case report. It helped to scientifically validate our own report. Maybe, in the future, someone would better reflect about the association between SJM and congenital heart diseases.

The authors.