CASE REPORT

A case of platypnoea orthodeoxia following Laproscopic Cholecystectomy

Timothy Riddles,¹ James Brown,² Daniel Judge³

¹Department of Thoracics, Cairns Hospital, Cairns, Queensland, Australia

²Department of Thoracic Medicine, Cairns Hospital, Cairns, Queensland, Australia

³Cairns and Hinterland Hospital and Health Service, Respiratory department, Cairns, Queensland, Australia

Correspondence to

Dr Timothy Riddles,
timothy.riddles@health.qld.gov.au

Accepted 27 March 2018

SUMMARY

Platypnoea Orthodeoxia syndrome is defined by dyspnoea and hypoxaemia exacerbated by orthostatic positioning and alleviated in recumbency. This condition has been reported in association with pulmonary, hepatic and cardiac disease. We describe a case herein of a 79 year old female who developed severe hypoxaemia and dyspnoea post laparoscopic cholecystectomy. A difference in arterial blood gas oxygen tension was demonstrated in the supine and erect positions on arterial blood gas analysis. A Patent Foramen Ovale (PFO) with a right to left shunt was evident on echocardiogram employing colour doppler and agitated normal saline studies. Definitive management, with closure of the patent foramen ovale lead to a complete resolution of symptoms and normalisation of arterial blood gas parameters in both the supine and erect positions.

BACKGROUND

This case highlights the importance of recognising Platypnoea Orthodeoxia (POS) syndrome as a cause for undifferentiated dyspnoea and without a high degree of suspicion, this may lead to a delay in diagnosis. This condition is likely to be under-reported. POS is a rare syndrome associated with a heterogeneous group of underlying pathologies. The syndrome can be extremely debilitating, with severe hypoxaemia.^{1–5} Those cases associated with an intra-cardiac shunt are readily amenable to definitive treatment with shunt closure.^{1,4,5} Where a PFO is the culprit anatomical defect, closure of the shunt may be performed using either an open surgical or a percutaneous approach. However, prior to shunt closure, right heart catheterisation should be obtained to exclude pulmonary hypertension. Failure to identify this prior to closure may lead to acute right heart failure with decompensation.¹ Diagnosis should be initially suspected with positional hypoxaemia and dyspnoea, and confirmed biochemically with arterial blood gas analysis.^{1,6}

CASE PRESENTATION

We describe a case of a 79 year old, otherwise well female presenting in type one respiratory failure following a recent laproscopic cholecystectomy for an impacted gallbladder neck stone. The post-operative course was complicated by; progressive dyspnoea on exertion, hypoxaemia with oxygen saturations on room air at 83%, and a single syncopal event precipitated by bending over.

During admission, progressive dyspnoea at rest prompted several emergency calls and a subsequent admission to the intensive care unit. The cause for her deterioration remained a diagnostic dilemma for a number of days and following a number of subspecialty reviews.

Examination revealed hypoxaemia with an oxyhaemoglobin saturation of 90% (table 1) despite oxygen supplementation via a non-rebreather mask, at fifteen litres per minute. Cardiovascular and pulmonary examinations were otherwise unremarkable with no evidence of pulmonary hypertension, congestive heart failure or pulmonary disease. It was noted that her oxygen saturations worsened with repositioning, this then prompted biochemical confirmation.

INVESTIGATIONS

Arterial blood gas analysis confirmed hypoxaemia, with a significant discrepancy between oxygen tension while supine compared with repeat analysis with upright positioning. Findings on arterial blood gas were consistent with a diagnosis for POS. A CT Pulmonary Angiogram (CTPA) initially suggested a sub-segmental pulmonary embolism; however, on review and with a repeat scan, there was no evidence of pulmonary arterial occlusion. A High-Resolution Computer Tomography (HRCT) scan of the chest demonstrated minimal atelectasis with mild emphysematous change and no other significant parenchymal pathology. Initial Trans-thoracic Echocardiography demonstrated a mildly dilated aortic root with mild aortic regurgitation and no flow across the intra-atrial septum was identified, however agitated saline was not administered and was performed supine. Further investigation with Transoesophageal Echocardiography (TOE) specifically looking for possible shunt showed a mildly dilated aortic root at 41 mm with moderate aortic regurgitation, mild segmental impairment of systolic function involving the inferior-lateral walls, and an intra-atrial septum bowed significantly to the left indicative of high right sided pressures. A Patent Foramen Ovale was observed and seen to be stretched and widely patent (figure 1), with a right to left shunt evident both with colour doppler and with agitated saline studies. A left heart catheterisation revealed mildly diffuse coronary artery disease and preserved ejection fraction by left ventriculogram of 55%. Right heart catheterization conclusively demonstrated a normal pulmonary artery pressure and confirmed a significant right to left shunt. The pulmonary to systemic blood flow

To cite: Riddles T, Brown J, Judge D. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-221714

Table 1 Change in oxygen tension with positioning pre and post PFO closure, as demonstrated with arterial blood gas analysis

Arterial blood gas analysis pre and post surgical closure	Positioning	
	Supine	Upright
Before PFO Closure		
pH	7.51	7.56
pCO ₂	26 mmHg	22 mmHg
pO ₂	56 mmHg	39 mmHg
Oxygen saturation	92 percent	83 percent
After PFO Closure		
pH		7.50
pCO ₂		32 mmHg
pO ₂		95 mmHg
Oxygen Saturation		98 percent

ratio (Qp:Qs) was measured at 0.7 supine and 0.5 while sitting, indicating both a right to left shunt, and of worsening shunt dynamics with erect positioning.

TREATMENT

The demonstration of dependent hypoxaemia with upright positioning was consistent with a diagnosis of POS. Investigations confirmed the underlying mechanism to be of a large right to left cardiac shunt through a moderate PFO. The patient subsequently underwent definitive repair of her PFO with a 25 mm PFO Amplatzer Occluder deployed via right heart catheterisation and under intra-cardiac ultrasound guidance. Repeat echocardiography revealed successful closure and repeat arterial blood gas analysis demonstrated correction of hypoxaemia. Symptoms of dyspnoea also resolved, consistent with the demonstrable improvement in oxygen tension (table 1).

OUTCOME AND FOLLOW-UP

Four years after, this female has continued to have a positive outcome from closure of her PFO. On latest outpatient review she remained free of platypnoea and dyspnoea, and arterial blood gas analysis and oxygen saturations still demonstrated normal oxygen tension.

DISCUSSION

POS has been reported in association with hepatic, pulmonary and cardiac conditions. The underlying pathophysiology is not adequately understood.¹ However, this syndrome is consistently

characterised by the combination of both an anatomical defect permitting a right to left shunt, as well as a functional component to promote shunting via either an elevation in right heart pressures or due to anatomical distortion favouring flow across the shunt.^{1-3 5 6} A number of intra-cardiac defects have been associated with POS. Defects across the atrial septum remain the most commonly identified, particularly the presence of a PFO.^{1,3-5} Whereas other cardiac causes, atrial septal defects, and a fenestrated atrial septum are more rare.⁶ Pulmonary arterio-venous malformations are also rarely associated with shunting in POS.⁶ Surgery is a commonly reported precipitant for the development of POS,^{1 3} and while various mechanisms have been proposed to cause a transient elevation in right heart pressures, the underlying mechanics that maintain the shunt are not well understood.³

The main drivers of physiological change during laparoscopy are from hypercarbia and elevated intra-abdominal pressure, both resulting from abdominal insufflation with carbon dioxide. Following initial insufflation, right atrial pressure increases following transient rise in venous return from compression of the splanchnic vasculature.^{7 8} Right atrial pressure elevation may be perpetuated by redistribution of portal venous blood to the central venous system through compression of hepatic arterial vessels, reduced portal venous flow to abdomen and with reduction in splanchnic blood flow.⁷ Cephalic displacement of the diaphragm from insufflation also increases atrial pressures and reduces pulmonary compliance.^{3 7} Right heart pressures may also be accentuated by hypercarbic alterations in vascular resistance, with both an increase in pulmonary arterial pressure and pulmonary vascular resistance.^{7 8} While these physiological changes may explain promotion of right to left shunting intra-operatively, the return to normal physiological state post operatively would assume reversal in the right to left shunt.

Possible explanations for mechanisms that maintain right to left shunting post-operatively remain speculative. A previous case of POS following laparoscopy was attributed to right diaphragmatic palsy.⁹ However, this was not evident in this case, with a chest radiograph demonstrating normal diaphragmatic position. Haemodynamic changes during laparoscopic surgery can increase myocardial oxygen demand resulting in ischaemia to the myocardium,⁷ and affecting right ventricular compliance. However, no compromise to right ventricular systolic function was reported on echocardiography. A common post-operative complication of atelectasis may increase pulmonary vascular resistance and cause a ventilation perfusion mismatch. While this patient did have evidence of atelectasis it is unlikely to be sufficient to explain this entirely. Another consideration is a peri-operative pulmonary embolism; however repeat CTPA did not demonstrate evidence for this. Given the physiological changes brought on by an elevation in intra-abdominal pressure from insufflation, it is possible that this patient's obesity contributed to sustained elevation in intra-abdominal pressure post-operatively and may have had a contributing role in maintaining shunt dynamics. However, one would expect this to be comparable with pre-surgical intra-abdominal pressures.

Low *et al* reported a similar case of POS following laparoscopic cholecystectomy.³ The mechanism proposed involved stretch of the intra-atrial septa resulting from the dilated aortic root, with erect positioning accentuating this stretch with possible compression on the right atria.³ The perioperative insult resulting in this was not clear. Perhaps the anatomical changes discussed by Low *et al* with horizontal shift to more horizontal axis of the intra-atrial septa, counter-clockwise rotation of the heart and mediastinal shift may be brought on permanently by

Figure 1 Transoesophageal Echocardiography demonstrating a widely patent foramen ovale and intra-atrial septum bowed to the left.

the temporary changes seen from insufflation. It is likely, that no single explanation can be entirely contributory.

We propose, similarly to others,⁵ that in this instance, an insult during surgery altered the underlying anatomy to yield a shift in cardiac axis, thus accentuating the functional mechanism of the shunt. The functional elements involved a combination of an enlarged aortic root, stretching of the intra-atrial septum and leading to a widening of the PFO. This promoted a change in atrial blood flow, directing blood across the intra-atrial septum. The peri-operative insult likely manifested from changes in intra-cardiac pressures, alteration in central venous flow, and increases in pulmonary vasculature resistance from laparoscopic surgery and which persisted either due to a permanent change in the underlying anatomy or cardiac axis, or due to a host of potential risk factors that permitted ongoing elevation in intra-abdominal pressure.

Patients' age has also been identified as a risk factor in the literature, with an over-representation of elderly patients diagnosed with POS.¹⁰ Previous studies have postulated an increase in age related ventricular compliance as a contributing factor.³ An additional mechanism may also be due to age related decline in connective tissue promoting anatomical distortion post operatively and change in cardiac axis leading to change in shunt dynamics.

Learning points

Platypnoea Orthodeoxia (POD) syndrome is:

- An uncommon cause of hypoxaemia in the inpatient population
- Likely to be underreported, and without a high degree of suspicion may lead to a delay in diagnosis
- Associated with a heterogeneous group of underlying pathologies
- Readily amenable to definitive treatment with shunt closure in cases associated with an intra-cardiac shunt

Contributors All listed authors have met the criteria for authorship set out in the ICMJE guidelines. All authors contributed to the thought, discussion, design and

editing of the report. The majority of the writing of the article was done by the lead author with contributions from co-authors. All authors have given their final approval prior to submission. Specifically, both Dr DJ and Dr JB were involved directly in care and management of the patient related to this case whilst in hospital, and have followed up with the patient as well through outpatient department. They were involved in diagnosis and coordination of management of this patient's care. Both Dr DJ and Dr JB have provided oversight and supervision in relation to the outline of the submitted case report. They have also contributed to revision and editing of draft copies of the case report as well. They have offered their opinions and guidance into the direction and content of the submitted article. Dr Riddles, as lead author was responsible for the majority of the writing of the document including development of tables and images. Dr TR was also responsible for coordination of submission of the article.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2018. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- 1 Blanche C, Noble S, Roffi M, *et al.* Platypnoea-orthodeoxia syndrome in the elderly treated by percutaneous patent foramen ovale closure: a case series and literature review. *Eur J Intern Med* 2013;24:813–7.
- 2 Vasant J, Jones S, Easaw J. Platypnoea-orthodeoxia syndrome. *BMJ Case Rep* 2011;2011:bcr1120103559.
- 3 Low Z, Darvall JN, Radford ST. Platypnoea-orthodeoxia syndrome post laparoscopic surgery in a patient with a patent foramen ovale. *Anaesth Intensive Care* 2014;42:385–8.
- 4 Nassif M, Lu H, Konings TC, *et al.* Platypnoea-orthodeoxia syndrome, an underdiagnosed cause of hypoxaemia: four cases and the possible underlying mechanisms. *Neth Heart J* 2015;23:539–45.
- 5 Buchholz S, Shakil A, Figtree GA, *et al.* Diagnosis and management of patent foramen ovale. *Postgrad Med J* 2012;88:217–25.
- 6 Knapper JT, Schultz J, Das G, *et al.* Cardiac platypnoea-orthodeoxia syndrome: an often unrecognized malady. *Clin Cardiol* 2014;37:645–9.
- 7 Atkinson TM, Giraud GD, Togioka BM, *et al.* Cardiovascular and Ventilatory Consequences of Laparoscopic Surgery. *Circulation* 2017;135:700–10.
- 8 Odeberg-Wernerman S. Laparoscopic surgery—effects on circulatory and respiratory physiology: an overview. *Eur J Surg Suppl* 2000;585:4–11.
- 9 Queirós C, Francisco E, Almeida J. Platypnoea-Orthodeoxia syndrome after complicated cholecystectomy: an unsuspected diagnosis. *Acta Med Port* 2017;30:827–30.
- 10 Richter S, Ohlow MA, Lauer B, *et al.* Platypnoea-orthodeoxia-syndrome in a 69-year-old male: a case report. *Clin Res Cardiol* 2008;97:402–6.

Copyright 2018 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow