Assessing potential divers with a history of congenital heart disease

Mark S Turner

Abstract

(Turner MS. Assessing potential divers with a history of congenital heart disease. 2015 June;44(2):111-115.)
This article describes a structured approach to assessing the medical fitness of potential divers who have a history of congenital heart disease. The importance of a complete and accurate cardiac history, including details of surgery and other interventions is emphasised. Specific assessment of intracardiac shunts, exercise capacity and ability to deal with the physical challenge of diving, risk of diving-induced pulmonary oedema, of arrhythmia and of incapacity in case of arrhythmia and the consequences of surgical and catheter treatment are discussed, including the risks associated with lung injury and the pressure limitations of implanted devices like pacemakers. Clinical assessment will usually include echocardiography and exercise testing with additional investigations such as MRI scanning, CT of heart or lungs, cardiopulmonary exercise testing and ECG monitoring, as required. Examples of different congenital lesions are given applying this approach (atrial septal defect, tetralogy of Fallot, bicuspid aortic valve and the Fontan circulation). The approach is based on an individual cardiologist's opinion and is not specifically evidence-based, but seeks to apply what is known in other areas of diving medicine to this potentially complex group of patients.

Key words
Fitness to dive; cardiovascular; pathology; physiology; children; right-to-left shunt; review article

Introduction

Due to the success of congenital cardiac surgery, interventional cardiology and paediatric intensive care, more than 85% of children with congenital heart disease reach adulthood. Furthermore, there are individuals who present in adulthood with congenital lesions.[1] There are now potential divers with congenital heart disease, some having had surgery or interventional treatment. There are no randomised controlled trials to guide us in this area, so this article is based on personal opinion and on an understanding of the mechanisms of diving illnesses and is influenced by the UK Sports Diving Medical Committee Guidelines.[2]

We aim to treat our patients so they can lead as normal a life as possible and there is good evidence that exercise improves patients with all forms of heart disease. Diving, however, carries specific risks that do not apply to most other sports, so special consideration of divers is justified. This article cannot be comprehensive and is not a replacement for an individualised specialist assessment; an accurate assessment of the 'patient' is needed, before the 'patient' can become a 'diver'. Table 1 summarises the key areas to consider when assessing potential divers.

Intracardiac shunts

'Hole in the heart' is a well-known form of congenital heart disease, such as atrial septal defect (ASD) or ventricular septal defect (VSD). ASD and VSD can be part of other types of congenital heart disease. VSD is part of Tetralogy of Fallot, but less well-known is that Ebstein's anomaly of the tricuspid valve is associated with an ASD or a large persistent foramen ovale (PFO) in 80% of patients.

In normal life, ASD will cause enlargement of the right ventricle, atrial arrhythmias, breathlessness and a small risk of stroke by paradoxical embolus (although the left atrial pressure is usually higher than the right). For divers, breathlessness and arrhythmia may be precipitated underwater, but perhaps more importantly paradoxical embolization of nitrogen bubbles during decompression causes some types of decompression illness (DCI). In a normal heart with PFO, the left atrial pressure is higher than the right, so the 'flap' is held down. The patient with Ebstein's anomaly, ASD, or tetralogy of Fallot is more likely to have a higher right atrial pressure, with a higher chance of right-to-left shunt, increasing the risks of DCI.

Obligate right-to-left shunts mean patients are desaturated at rest. These are patients with complex circulations with mixing of the systemic venous (blue) blood with the pulmonary venous (red, oxygenated) blood. Therefore, measuring oxygen saturation is important for congenital heart disease patients. Other lesions can cause desaturation, such as a left superior vena cava draining straight to the left atrium, where bubble contrast echocardiography (echo)

Table 1
Issues for assessment in patients with congenital heart disease

- Potential for right-to-left shunt;
- Ability to deal with the physical challenges of diving;
- Risk of diving-induced pulmonary oedema;
- Risk of arrhythmia/sudden loss of capacity;
- Risk of non-cardiac issues/associations; e.g., lung damage post thoracotomy, bullae in Marfan syndrome;
- Drug therapy/pacemaker.

performed from the left arm, will cause the left atrium to fill with many bubbles, whereas a bubble echo from the right arm, could be falsely negative. This underlines the need to understand the cardiac anatomy.

Assessment of right-to-left shunt

Given the high prevalence of right-to-left shunt in patients with congenital heart disease, bubble contrast echocardiography, according to the protocol developed by Wilmshurst et al,[3] should be performed in all potential divers to assess the risk of DCI; and the magnitude of the shunt determines the risk.[3] The bubble echo must be performed in accordance with this protocol and the images should be quality controlled. The bubble images must show excellent opacification of the right heart, with complete filling with microbubbles. If gaps exist between the bubbles, then opacification is inadequate and can underestimate the shunt. Furthermore the quality of the provocative manoeuvres must be assessed. With a good Valsalva, it should be possible to see the left-sided heart chambers become smaller during the Valsalva, and recover their size after release. The image must not be interrupted at the time of Valsalva release, maintaining a view of the left ventricle, as the bubble shunt can sometimes only be seen for a few beats. No medical test is perfect, but bubble contrast echo is very operator-dependent, and sadly many studies sent to us from elsewhere fail our quality control criteria and can be falsely negative.

For those with moderate or large shunts, diving may still be possible but with very careful management of gas load. If the gas load is low, venous bubbles do not form, and the tissues do not build up a significant partial pressure of inert gas. As the pathophysiology of shunt-related DCI relies on venous bubbles being present to cross the PFO, or other defect, and then to embolise to tissues where there is already a significant inert gas burden, modification of gas load, and avoiding lifting or straining after surfacing should effectively reduce the risk. The UKSDMC recommends 15 metres, or equivalent air depth or the use of the DCIEM tables as suitable risk-reduction measures (although this may not be applicable to less fit or older individuals with underlying cardiac pathology).

For individuals with very large or resting shunts, and especially those with desaturation, my view is that they must be at higher risk of DCI and may not be fit to dive, or may need more rigorous limitation of gas load. If the desaturation is because of complex pathology, there may be other reasons to be unfit for diving.

Ability to deal with the physical challenges of diving

Some patients with congenital heart disease can achieve normal cardiopulmonary performance on exercise testing, whereas others will always be limited. Increasingly we use cardiopulmonary testing clinically, so this information may be available. During diving the cardiac output may need to increase to deal with exercise or anxiety, or to undertake safety-critical tasks. Good ventricular function and well-functioning valves on echocardiography or cardiac MRI scanning is reassuring, especially if accompanied by a normal cardiopulmonary exercise test.

Patients with common lesions such as repaired tetralogy of Fallot have a wide spectrum of exercise capacity and symptoms. Some will have severe pulmonary regurgitation and require early pulmonary valve replacement, whereas others can survive after primary repair with good function and do not require repeat surgery.[4] Thus, careful clinical assessment of the individual coupled with MRI and cardiopulmonary exercise testing is usually needed.

Immersion pulmonary oedema (IPE)

Ventricular function, valve function and exercise capacity are also relevant to diving-induced IPE. In IPE's 'purest' form, the systemic vascular resistance increases pathologically during immersion, due to an exuberant vasoconstriction, making the heart pump against a greater resistance. This in turn increases the left atrial pressure, such that the pulmonary capillaries leak and pulmonary oedema commences.[5] If the left atrial pressure starts high due to impaired ventricular or valve function, or hypertension, left atrial pressure has less to rise before the onset of pulmonary oedema. As some vasoconstriction to cold and immersion is normal, and the compression of abdomen and legs may also increase atrial pressures, a high starting left atrial pressure is likely to put the diver at higher risk of IPE. Furthermore, a diseased ventricle will need to increase the left atrial pressure more than usual in order to increase cardiac output, as increasing contractility may be limited by the underlying ventricular disease. Tissue Doppler measurements may estimate left atrial pressure, and should be performed during a transthoracic echocardiogram. Severe limitation to exercise capacity, significant ventricular impairment, evidence of elevated left atrial pressure, or significant valve disease are probably barriers to diving, but may be an indication for corrective surgery or intervention.

Conditions severely limiting the increase in cardiac output with exercise

Aortic stenosis, pulmonary stenosis, pulmonary vascular disease and hypertrophic obstructive cardiomyopathy can cause an inability to increase the cardiac output that is so profound that collapse can occur on exercise. Any potential diver with a history of exercise-related collapse is at markedly increased risk and should not dive until the underlying cause is treated, and they are reassessed. Symptomatic patients with hypertrophic cardiomyopathy are unlikely to be candidates for diving and our knowledge of diving risk in asymptomatic patients with hypertrophic cardiomyopathy is inadequate to make any recommendation.

Arrhythmias

Arrhythmias are the commonest reason for emergency admission in adult congenital heart patients, atrial arrhythmia being the most common; however, ventricular arrhythmia is more likely to be incapacitating and life threatening.[1] For this reason, any patient who has had ventricular arrhythmia without a definite and reversible cause, is likely to be at greater risk and is unlikely to be fit to dive.

Atrial arrhythmias can cause haemodynamic compromise, if the rate is very fast, or if the underlying heart or valve function is poor. Thus, if the diver is on rate-controlling medication and the heart function and exercise capacity are good, the diver is unlikely to come to harm if they suffer an atrial arrhythmia, so long as they can surface immediately (without obligatory decompression stops). How the diver feels when they get atrial arrhythmia on the surface is a key part of the history in this situation.

Consequences of treatment/associations

To survive, many patients will have had surgery or catheter-based interventions which could have an impact on diving fitness. Previously arterial shunts (e.g., Blalock-Taussig) were often used to increase pulmonary blood flow for cyanotic patients, e.g., tetralogy of Fallot. Modern surgery aims for a primary repair at a younger age, but potential divers may have been treated with shunts, which are usually performed through a thoracotomy. Thoracotomy has a potential to cause lung damage and scoliosis. A further risk to lung function is that phrenic nerve palsy can occur during complex congenital heart repairs. These lesions may be risk factors for pulmonary barotrauma, may compromise lung function and potentially limit exercise capacity.

Catheter interventions should not damage the lungs, but pneumothorax at the time of subclavian puncture (such as for pacemaker implantation or lines for intensive care or dialysis) could occur and the potential diver's history needs to be scrutinised for this type of complication. A traumatic pneumothorax from needle injury may not increase the risk of pulmonary barotrauma, but lung or pleural scars from infections or pleural adhesions may have an impact.

Pacemakers are no longer just used to treat bradycardia and so the mere presence of a pacemaker is not enough information to inform decisions about fitness to dive. Patients with cardiac resynchronisation therapy (CRT) will have had an impaired left ventricle and heart failure symptoms – they will, therefore, have reduced exercise capacity and increased risk of IPE unless they have responded dramatically to the therapy. Implantable defibrillators are used for those at very high risk of ventricular arrhythmia, so are unlikely to be fit to dive. Some defibrillators still function normally at 709 kPa (St Jude Medical USA),[6] but it is the underlying heart disease that is the contraindication to diving. If the risk of arrhythmia is not as high, e.g., if the ICD was implanted for primary prevention, or if there was a clear precipitant, one may make an individual decision about diving fitness. In these cases, mitigation of the risk of arrhythmia, or ICD discharge may be appropriate, such as the use of a full-face mask that would prevent inhalation of sea water in case of blackout, diving with an experienced buddy and avoiding obligatory decompression stops. Furthermore, different ICDs and pacemakers are tested to different pressures, so the device must be precisely identified.[7] For example, most St Jude Medical devices are tested to 7 ATA (709 kPa), whereas Medtronic are tested to 2.5 ATA (253 kPa), and devices may malfunction if taken beyond their tested depths.

Clinical assessment

PAST HISTORY

It is essential to obtain a detailed past medical history, including the nature of surgical and/or other interventions, their complications, other medical problems and a full drug history. Understanding the detail, such as thoracotomy or sternotomy, is important when assessing risk and potential lung damage. Remember to discuss other congenital abnormalities and whether the cardiac lesion is part of a syndrome. Ask about exercise capacity and exercise-related symptoms, palpitations, migraine with or without aura, blackouts and collapse.

EXAMINATION and ECG

Examine for blood pressure, murmurs, scars (thoracotomy, sternotomy and groin scars) and for the presence of a pacemaker, which may be in a pectoral position or could be in the abdomen. The morphology of the QRS complexes and T-waves may be abnormal, so an abnormal ECG in itself may not exclude the potential candidate. However, recording an ECG is important to assess the cardiac rhythm and to assess conduction abnormalities. Ambulatory ECG monitoring may be needed if there is an increased risk of arrhythmia or heart block.

ECHOCARDIOGRAPHY

Echocardiography is important to identify and quantify valve lesions, assess ventricular function and look for other congenital cardiac abnormalities such as coarctation of the aorta. So, a supra-sternal view is needed, which is not performed by all echocardiographers, even though it is part of a standard examination performed in high-quality departments. Bubble contrast echocardiography will often be needed, and should include the use of provocation manoeuvres to promote right-to-left shunt such as Valsalva release and sniffing.[3] Many departments perform bubble contrast echocardiography in a way that does not satisfy our quality control criteria, so the rigour with which this test is performed is extremely important if false negative tests are to be avoided.

As described earlier, bubble contrast should totally opacify the right heart so, if any 'black', unopacified space can be seen in the right heart, the contrast is inadequate (bubbles are white and should not be separated by any blackness at all). The bubbles in the right heart need to be present at the time of a sniff or Valsalva release, the left heart should get noticeably smaller with the Valsalva (as the venous return is restricted), the septum should bulge to the left on Valsalva release and bubbles should be seen along the entire length of the septum as it bulges. The echo pictures need to be recorded for many beats – I usually record at least 10 beats – so that the pattern and timing of bubbles entering the left heart can be seen. Any scans not satisfying these criteria cannot be used for risk assessment, and need to be repeated. Transoesophageal echo is very rarely needed and is less sensitive for shunts, although many echocardiologists who are not PFO specialists will disagree with this.

EXERCISE TESTING

Exercise testing can be used to confirm that there are no exercise-induced arrhythmias, can give some reassurance about ischaemia (but more sensitive ischaemia tests are now available if ischaemia is a specific issue, e.g., for patients after the arterial switch operation where the coronaries are moved) and gives reassurance that the cardiac output can be increased. Tests such as cardiopulmonary testing or the Chester step test[8] can give reassurance about exercise performance and provide some reassurance about IPE, although IPE can occur even in very fit individuals if the pathophysiology is solely extreme vasoconstriction. ECG monitoring may be appropriate if there is concern over arrhythmia or intermittent heart block.

OTHER TESTS

Spirometry and chest radiography are indicated in all potential divers who have had chest surgery (thoracotomy or sternotomy) to help to determine if there is any lung damage that could increase the risk of pulmonary barotrauma, although normal lung function and chest radiography cannot exclude this risk. Some cardiac abnormalities may be associated with lung abnormalities, and if there is a high index of suspicion, chest CT scanning may be needed to identify bullae, scarring or other abnormalities.

Four clinical examples of the systematic approach to assessment are presented in Table 2.

Summary

This article has attempted to provide a framework for assessing potential divers with a history of congenital heart disease. It does not replace a full assessment by a (congenitally-trained) cardiologist with experience in diving medicine, but seeks to illustrate some of the issues and complexities that need to be evaluated. The overwhelming

Table 2

Four case examples of the systematic approach to assessing potential divers with a history of congenital heart disease

1. Secundum ASD

Intracardiac shunt
Untreated: increased risk of DCI;
Treated: bubble echo for residual shunts;
Cardiac function and exercise capacity
Untreated or treated: assess ventricular size and function, right ventricular size, right ventricular pressure and exercise capacity;
Risk of IPE: likely to be low if ventricular function and exercise capacity are okay;
Arrhythmia risk: age and pathology dependent; risk of atrial fibrillation (usually well tolerated).

2. Tetralogy of Fallot

Unoperated ToF
Patient should not dive as they have obligate right-to-left shunts, poor cardiopulmonary exercise performance and systemic right ventricular pressures.
Operated ToF patients
Right to left shunt: assume PFO or ASD; undertake bubble contrast echo; residual ventricular septal defect (VSD) may be present, and if the right ventricular pressure is elevated (as it commonly is) then right to left shunt across the VSD may be seen, but should also be identified with bubble contrast echocardiography.
Cardiac function and exercise capacity: pulmonary regurgitation (PR) and right ventricular dilatation common; exercise capacity should be normal or nearly normal.
Risk of IPE: likely to be low if ventricular function and exercise capacity are okay;
Consequences of surgery: impaired lung function common (previous thoracotomy, scoliosis, potential for lung hypoplasia due to poor pulmonary flow in early life);
Arrhythmia risk: both atrial fibrillation and flutter can occur, but also ventricular tachycardia (most likely to cause collapse).

3. Bicuspid aortic valve

Right to left shunt: association with PFO, so bubble contrast echo indicated;
Ability to exercise: depends on severity of valve dysfunction and ventricular function;
IPE risk: depends on severity of valve lesions, ventricular function, coexistent coarctation of the aorta;
Arrhyhmia risk: atrial fibrillation risk, which can cause collapse if valve lesions severe or ventricular function impaired;
Associations: aortic enlargement (aortopathy) and dissection risk present.

4. Fontan circulation

This is a single ventricle repair, with non-pulsatile flow to the lungs, and is used only for severe forms of heart disease where a two ventricle repair is not possible.
Right to left shunt: very common, can be at atrial level or via venous collaterals;
Exercise capacity: low fitness levels and poor cardiopulmonary exercise tests;
IPE: the risk of IPE hard to predict, but vasoconstriction would cause low cardiac output;
Arrhythmia: atrial arrhythmia is common in Fontan patients, often causing haemodynamic compromise on the surface.

principle is that the underlying cardiac condition needs to be defined as accurately as possible to allow a meaningful risk assessment. Once additional risks are understood, the diver can mitigate the risks such as by avoiding decompression obligations, diving with experienced buddies and avoiding remote or extreme diving.

Furthermore, while some of the recommended testing may reassure us that there is not an excessive risk associated with diving, any patient with a congenital heart disease history is likely to be at a slightly greater risk than a similarly aged diver without any cardiac history. Therefore, a discussion with the individual is needed to ensure that they are fully informed of additional risks that diving may pose. For this reason, I would be reluctant to assess a child with congenital heart disease for diving, as my personal view is that the individual needs to 'own' this unquantifiable risk themselves, rather than their parents taking responsibility for it.

References

1 Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, et al. ESC guidelines for the management of grown-up congenital heart disease (new version 2010). Task Force on the management of grown-up congenital heart disease of the European Society of Cardiology (ESC); Association for European Paediatric Cardiology (AEPC); ESC Committee for practice guidelines (CPG). *Eur Heart J*. 2010;31:2915-57.

2 *United Kingdom Sports Diving Medical Committee Medical Standards*. [cited 2015, April 10]. Available from: http://www.uksdmc.co.uk/index.php?option=com_content&view=section&id=1&Itemid=2

3 Wilmshurst PT, Byrne JC, Webb-Peploe MM. Relation between interatrial shunts and decompression sickness in divers. *Lancet*. 1989;334:1302-6.

4 Frigiola A, Hughes M, Turner M, Taylor A, Marek J, Giardini A, et al. Physiological and phenotypic characteristics of late survivors of tetralogy of Fallot repair who are free from pulmonary valve replacement. *Circulation*. 2013;128:1861-8.

5 Wilmshurst PT, Nuri M, Crowther A, Webb-Peploe MM. Cold-induced pulmonary oedema in scuba divers and swimmers and subsequent development of hypertension. *Lancet*. 1989;333:62-5.

6 *Effects of scuba diving and hyperbaric oxygen therapy on St. Jude Medical implantable cardiac rhythm devices*. [cited 2015, April 10]. Available from: http://professional-intl.sjm.com/resources/emi/dental-medical

7 Kot J. Medical devices and procedures in the hyperbaric chamber. *Diving Hyperb Med*. 2014;44:223-7.

8 Sykes K, Roberts A. The Chester step test – a simple yet effective tool for the prediction of aerobic capacity. *Physiotherapy*. 2004;90:183-8.

Acknowledgements

Dr Turner would like to acknowledge the support of his colleagues in the UK Sports Diving Medical Committee (UKSDMC), for discussion of patients/divers. In particular, Dr Peter Wilmshurst has generously given of his time and expertise to help in decision-making about patients with complex heart disease. Dr Turner would like to express thanks to the UKSDMC for an educational grant to support his attendance at the South Pacific Underwater Medicine Society Annual Scientific Meeting in 2014, when the presentation leading to this article was made.

Conflicts of interest

Dr Turner is a Consultant and Proctor for St Jude Medical (structural heart division), Medtronic Inc and Edwards Lifesciences (heart valve divisions) and a Consultant and Lecturer for Gore Medical.

Submitted: 13 April 2015
Accepted: 01 May 2015

Mark S Turner, Bristol Heart Institute, Bristol, UK

Address for correspondence:
Level 7 Bristol Heart Institute
Bristol Royal Infirmary
Marlborough Street
Bristol BS2 8HW
United Kingdom
E-mail: *<markturner3@nhs.net>*

The database of randomised controlled trials in hyperbaric medicine maintained by Michael Bennett and his colleagues at the Prince of Wales Hospital Diving and Hyperbaric Medicine Unit, Sydney is at:
<http://hboevidence.unsw.wikispaces.net/>

Assistance from interested physicians in preparing critical appraisals is welcomed, indeed needed, as there is a considerable backlog. Guidance on completing a CAT is provided. Contact Associate Professor Michael Bennett: <m.bennett@unsw.edu.au>