Reviewer’s report

Title: Single Ventricle with Persistent Truncus Arteriosus: A Case Report of Two Rare Entities in an Adult Patient

Version: 2 Date: 2 August 2007

Reviewer: Leisa J Freeman

I am familiar with the literature and believe that this case meets one of the 7 criteria for evaluation in the journal: New associations or variations in disease processes

Has the case been reported coherently?: Yes

Is the case report authentic?: Yes

Is this case worth reporting?: Yes

Is the case report persuasive?: No

Does the case report have explanatory value?: Yes

Does the case report have diagnostic value?: Yes

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

Comments to authors:

General

Revisions necessary for publication

The first paragraph is too staccato in places and wordy in others. It could be smoothed. Although the FH was ‘negative for congenital defects’ it would be good to state if 22q11 deletion was or was not sought in this patient at any time during his life. Did this patient have children?

Figure 1 (ECG) was not included. There is no description of the ECG either in the text (page 4) or below the absent figure 1 though a page is available. Is it thought that this was a sinus tachycardia or some form of IART?

Figure 2 (echo) no legend i.e this is a 4 chamber view etc

It is a shame that there is no post mortem photograph – was there one taken – if so it would be very helpful. Was the right lung thought to be hypertensive. The coronary arteries are said to be normal but often the LAD is small and the conus
branch of RCA is large which is important for the surgeons to know at op.
Truncus often has skeletal, ureteric and bowel abnormalities – did he??

It is helpful to explain the types of SV and common arterial trunk for education and the early paediatric survival etc BUT this case survived to adulthood and the authors should expand their reasoning for his survival i.e. the pulmonary trunk stenosis (which in itself is rare) was sufficient to allow survival into adult life which has been previously reported in Truncus arteriosus and prevented early death from heart failure. This then explains the benefit from the BT shunt. However it would be useful with the retrospectoscope to comment that arterial shunts now are not regarded favourably since they lead to further volume loading of the systemic ventricle as may be considered in this case. Given the admission aged 27 with a diagnosis of thrombosis of BT shunt they should consider whether the learning point should have been to continue oral anticoagulation rather than just aspirin after admission age 27? Given his extreme state on his terminal admission could they speculate that he should have had thrombolysis eg urokinase or tenecteplase and whether that would have altered his outcome? Was he given too much fluid given his apparent heart failure?

They could elucidate what intervention might have been proposed if he had survived. (eg truncal valve repair/replacement and tcpc) This would be helpful for the adult community if they were to be presented with such a case.

His arrhythmia management with propanenone might also be commented upon since the evidence of heart failure would suggest impaired ventricular function which might lead to avoidance of this drug. The authors should highlight the fact that new arrhythmias in patients with congenital heart disease should always trigger a search for haemodynamic change before just treating with an antiarrhythmic as was the case when he had VT age 35.

They note the only other case survived with this condition was what was called type 4 truncus (i.e pulmonary atresia) (ref 4) which in van praagh classification usually has interrupted arch or severe coarctation – did it? Type 4 of Collett and Edwards is now thought to be a variation of pulmonary atresia and large VSD – rather than considered in the spectrum of common arterial trunk.

Conclusion:
This case report is important but needs more relevance as to why it survived and what learning points about management of adults with congenital heart disease could be learnt along the way. This might also include a comment on the move away from venesection when heamatocrit is <65%. We are not told if the patient was iron deficient which would also have exacerbated his problems and this could be highlighted in the discussion.

**What next?:** Revise and resubmit

**Quality of written English:** Acceptable