1. Between 40-60% of babies with Down syndrome have congenital heart defects.\textsuperscript{1-3} Of these, 30-40% have complete atrioventricular septal defects (AVSD).\textsuperscript{2-5} Most AVSDs can be successfully treated if the diagnosis is made early and the baby referred for full corrective surgery before irreversible pulmonary vascular disease (PVD) is established.\textsuperscript{6-14}

2. There must be a high level of clinical suspicion of congenital heart disease for all newborns with the syndrome.\textsuperscript{8,15} Those diagnosed in the early neonatal period should be referred for urgent cardiac assessment as outlined below. Babies diagnosed in the later neonatal period should have accelerated referral to a paediatric cardiologist or a paediatrician with appropriate paediatric cardiological training especially if the 6 week deadline has been exceeded.

3. It is highly desirable to establish the cardiac status of every child with Down syndrome by age 6 weeks.\textsuperscript{5,13-15} Irreversible PVD is more likely to develop quickly in children with Down syndrome.\textsuperscript{2,9,10,13,16} Ideally surgery for AVSD is desirable by 6 months of age.\textsuperscript{9,13,17} Early cardiac surgery is desirable for children with Down syndrome with surgically correctable cardiac lesions to optimise outcomes.\textsuperscript{5-12,17}

4. Clinical examination alone is insufficient to detect even some of the most serious abnormalities.\textsuperscript{4,8,15,18}

5. It is very unlikely that a serious abnormality requiring early intervention (e.g. AVSD) will be missed if the following course of action is taken.\textsuperscript{4,8,15,19}

   - Clinical examination, electrocardiogram (ECG) and echocardiogram (ECHO) performed by someone with appropriate paediatric cardiological training\textsuperscript{5,8} for all newborns with Down syndrome ideally by the age of 6 weeks (in particular those with a superior QRS axis on ECG).\textsuperscript{20}
   - Telemedicine, if available, may provide a useful intermediate step between paediatrician and cardiologist.
   - Those with suspected problems should be referred for immediate paediatric cardiological review so that intervention, if necessary, can take place before pulmonary vascular disease develops.

   **Fetal Echocardiography**
   Given the absence of a uniform standard of fetal echocardiography we suggest that those who have had a fetal ECHO should still follow the above neonatal pathway.
   Older children who have never had an echocardiogram
   - Those with a normal ECG, no symptoms and no abnormal clinical signs should be referred routinely for additional assessment by a clinician with appropriate paediatric cardiological training.
   - Those with symptoms and/or abnormal clinical signs or ECG abnormalities should be referred urgently for assessment

6. People with Down syndrome with heart lesions are at increased risk of infective endocarditis.\textsuperscript{21 to 24} Therefore, parents and carers of all children with Down syndrome with heart lesions should be given verbal and written information about infective endocarditis preventive measures.\textsuperscript{25-27}
7. It should be remembered that despite a normal echo at birth children with Down syndrome, like all other children, can develop symptoms and signs of heart disease at a later age e.g. secondary to airway/respiratory problems.\textsuperscript{13, 28, 29} and may be at increased risk of developing pulmonary vascular disease and right heart failure.

8. Echocardiography may occasionally fail to diagnose AVSD and other major cardiac lesions, particularly in the first few days after birth, even when undertaken by skilled practitioners.\textsuperscript{8} As a result, if symptoms or signs of cardiac disease are detected at any age even where the early ECHO has been reported as normal, there should be a low threshold for repeating the ECHO examination.

9. There is an increased incidence of mitral valve prolapse (MVP) and aortic regurgitation (AR) from late adolescence into adulthood in people with Down syndrome which may be asymptomatic.\textsuperscript{22, 23,30-33} This has implications for infective endocarditis prevention particularly because of the high incidence of periodontal disease among this population.\textsuperscript{21--27} We therefore recommend careful cardiac evaluation including echocardiography for all people with Down syndrome early in adult life.\textsuperscript{8,30, 33, 34}

10. MVP occasionally progresses to mitral valve regurgitation (MVR). We recommend monitoring for signs of atrial fibrillation and/or left ventricular failure\textsuperscript{30, 31} in these patients, and some may be advised regarding restriction of competitive sporting activities.\textsuperscript{31,3}

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