DOWN SYNDROME : CONGENITAL HEART DISEASE

Key Points

(taken from 'Recognising heart disease in children with Down syndrome'. Archer et al. In Press.)

- Around 397 babies with Down syndrome and congenital heart disease are born every year in the UK, of whom 139 can be expected to have AVSD

- Surgical intervention for major shunt lesions must take place in early infancy to ensure optimum outcome hence the cardiac status of every baby with Down syndrome should be established and where appropriate an action plan agreed by age 6 weeks (see guideline for details)

- Without surgical intervention the outcome of AVSD is significant cardiac disability and premature death

- Clinical examination alone is insufficient to detect all of even the most serious heart disease in newborns and young babies

- All newborns with the syndrome in addition to careful clinical examination should have either neonatal echocardiography or ECG

- Neonatal echocardiography is undoubtedly the most effective single diagnostic procedure but it is not 100% effective in the early neonatal period and has drawbacks as well as resource implications

- Superior QRS axis on ECG is highly predictive of AVSD and such babies should proceed to urgent echocardiography

- Every child health team needs to have an agreed screening protocol in place to ensure the early diagnosis of serious heart disease.