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Management of pulmonary hypertension in Down syndrome

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The incidence of congenital heart disease CHD in Down Syndrome (DS) is about 40%. Many will have large septal defects. Previously these were often unrecognised and untreated leading to Eisenmenger syndrome. In addition, there are many other reasons for pulmonary hypertension PH in DS.
The pulmonary hypertension clinic at the Bristol Heart Institute is responsible for a population of around 6.5million. A total of 208 patients have been seen in this service since its inception in 2005, 59 with DS. Transition arrangements cover the transfer from our paediatric clinic to the adult clinic. Shared care arrangements with the Hammersmith hospital have led to the clinic being performed jointly in Bristol.
Before 2005, many patients with DS were unable to access therapy due to the excessive journey times; the furthest location being 222 miles away, representing a 14 hour round trip or journey by fixed wing aircraft. Such a journey to travel up to London in addition was often prohibitive for a patient with learning difficulties and their carers.
Of the 59 patients with DS, with mean age 28 years, 17 were male. All had CHD, 8 having had palliative cardiac surgery, the rest being untreated. At start of therapy, mean oxygen saturations were 65% and mean six minute walk distance (6MWD) was 120m.Treatment was with oral medication in 28 patients for an average of 1.7 years. Intravenous therapy was not deemed to be appropriate in any patient and one received inhaled Iloprost. Oral sildenafil was avoided in male patients with DS due to the side effects. 17 received Bosentan, 13 received sildenafil (4 having dual therapy) and one received Ambrisentan. Others were either too well (NYHA class II) or too stable to want therapy.
There were no complications of therapy. None had abnormal liver function tests, 6MWD increased after therapy by a mean of 45m and quality of life was significantly improved on questionnaire (CAMPHOR). Only 7 patients died in the 6 years of the clinic.
This early data represents benefits of therapy for a specific group of patients previously denied access because of their special needs.