Abstract Information

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Abstract Content **100%**

The TOPP registry was designed to provide current demographic, diagnostic, clinical and outcome data in pediatric PH. One primary objective was to describe the current work up/testing in pediatric practices to diagnose PH. Patients diagnosed with PH between 3 months and 18 yrs were eligible for enrollment. Investigators reported ECG, chest X-ray(CXR), echocardiography (ECHO), and Holters were done and results as 'normal' or 'abnormal.'

Between Jan 2008 and Feb 2010, 456 patients were enrolled. The majority of patients had ECGs, ECHOs and/or CXR performed: ECG (n=430, 94%; 90% 'abnormal'), ECHO (n=439, 96%; 99%'abnormal') and CXR (n=406, 89%; 80% 'abnormal'). None of the patients had normal results for all 3 tests, although2 tests were normal in 3% and 1 was normal in 18% . Additional tests included Holter(n=94, 21%; 56% 'abnormal'), routine laboratory studies (n=411, 90%), BNP (n=97, 21%) or NTproBNP (n=95,21%), cardiopulmonary exercise testing (n=34, 7%, 91% ≥ 7 years), 6-minute walk test (n=175, 38%), pulmonary function tests (PFT) (n=122, 27%), overnight oxygen saturation/sleep study (n=129, 28%), lung scan (n=104, 23%, 41% 'abnormal'),pulmonary angiography (n=198, 43%, 43% 'abnormal'), chest CT (n=189, 41%, 74%'abnormal'), magnetic resonance imaging (MRI) (n=42, 9%, 83% 'abnormal') andlung biopsy (n=21, 5%, 90% 'abnormal'). Exercise tests were performed most often in patients >7yrs($p<0.0001$). The analysis of age group distribution of the tests shows that test requiring patient cooperation are seldom performed below 7 years of age. No changes were observed in the tests performed through the 10 years during which the patients were diagnosed suggesting no significant changes in thediagnostic approach during this 10 year period (2001-2009). Seriouscomplication during cardiac catheterization were reported in 6% of the patients including hypotension(46%), arrhythmias(15%) pulmonary hypertensive crisis (19%) and unexpected intensive care admission (15%) . No deaths during catheterization were reported.

ECHO, ECG, and CXR were the non-invasive tests most frequently used to evaluate patients with suspected PH. At least one of these tests was abnormal in all patients with confirmedPH , suggesting that they can be used collectively to screen for suspected PH.A complete work-up was not performed in all patients. The use of some tests is dictated by age and cooperation. The low rate of exercise testing can be partly explained by the inability of children <7 years to perform reliable testing.It remains to be determined if this more limited approach is appropriate to discriminate different PH etiologies.