

**Diagnostic Evaluation in Pediatric Pulmonary Hypertension (PH) - Results from the First Pediatric Global Registry: Tracking Outcomes and Practice in Pediatric Pulmonary Hypertension (TOPP)**

*Beghetti M.(1), Berger.R.M.F.(2), Schulze-Neick I.(3), Raskob G.(4), Day R.(5), Pulido T.(6), Feinstein J.(7), Barst R.J.(8), Humpl T. (9)*

*1 Pediatrics, Hôpital des Enfants, Geneva, Switzerland; 2 Pediatrics, University Medical Center Groningen, Netherlands; 3 Pediatrics, Great Ormond Street Hospital for Children, London, UK; 4 University of Oklahoma, Oklahoma, US; 5 University of Utah, Salt Lake City, US; 6 National Heart Institute, Mexico City, Mexico; 7 Stanford University Medical Center, Palo Alto, US; 8 Pediatrics, Columbia University, New York, US; 9 Pediatrics, University of Toronto, Toronto, Ontario, Canada .*

**Introduction**

The TOPP registry (31 sites, 20 countries) 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 mos 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.'

**Methods and Results**

Between Jan 2008 and Feb 2010, 456 patients were enrolled. RHC confirmed PH in 435 (95%); in the remaining 5%, the diagnosis was made by pathology or echo. 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 of these tests, although 2 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%  $\geq$  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') and lung biopsy (n=21, 5%, 90% 'abnormal'). Exercise tests were performed most often in patients  $>7$  yrs( $p<0.0001$ ).

**Conclusions**

ECHO, ECG, and CXR were the non-invasive tests most commonly used to evaluate patients. At least one of these tests was abnormal in all patients with PH confirmed by RHC, suggesting that they can be used collectively to identify affected patients. A complete work-up was not performed in all patients. 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.