Scoliosis on measured height. Standard deviation score calculated from the Global Lung Initiative (GLI 2012) data was used to identify change.

Demographic data including date of birth, postcode, gender, date of surgery, height prior to surgery, Cobb angle, and the vertebrae involved in surgery was collected.

Results Children with scoliosis have reduced forced expiratory volume in 1 second (FEV1) and reduced forced vital capacity (FVC), with median Z scores of -1.5 and -1.1 respectively. Lung function undertaken between two and three years after surgery showed an absolute improvement, but no change in Z score, suggesting some lung function may be permanently lost despite the skeletal correction.

Conclusion Scoliosis surgery can halt the decline in lung function but does not result in improved lung function at 1–3 years post-surgery.

Under pressure: an update in pulmonary vascular disease

PREDICTING POSTCAPILLARY PULMONARY HYPERTENSION: VALIDATION OF THE H2FPEF AND OPTICS SCORES

H Stubbs, MK Johnson. Scottish Pulmonary Vascular Unit, Glasgow, UK

Background Distinguishing pulmonary arterial hypertension (PAH) from postcapillary pulmonary hypertension (PH) is crucial yet can be challenging. The H2FPEF and OPTICS scores have been proposed as predictors of an elevated pulmonary artery wedge pressure, in order to inform whether to proceed with further investigations for PH, including right heart catheterisation. These scores include routinely available information including age, comorbidities and transthoracic echocardiogram and electrocardiogram indices. The aim of this study was to externally validate the H2FPEF and OPTICS scores for use in vetting new PH referrals.

Methods A retrospective analysis of was undertaken of all patients who were referred to a tertiary PH centre in Scotland between 2016 and 2020. Patients were included if they have undergone diagnostic admission for PH, including right heart catheterisation, and were subsequently diagnosed with idiopathic PAH, heritable PAH, pulmonary veno-occlusive disease or postcapillary PH. Records were screened for components of the scores, which were calculated for each patient and compared to the post-investigation diagnosis as judged by multidisciplinary consensus. A H2FPEF score of ≥6 and an OPTICS score of ≥104 were used as thresholds for predicting postcapillary PH.

Results 107 patients with precapillary pulmonary hypertension and 86 patients with postcapillary pulmonary hypertension were included. Retrospective application of the OPTICS score demonstrated that pre-test scoring would detect 28% of cases with postcapillary pulmonary hypertension (sensitivity 0.28) yet at the cost of misdiagnosing 4% of patients with PAH as postcapillary PH (specificity 0.96). The H2FPEF score had a far greater sensitivity (0.70) yet reduced specificity (0.91), implying 9% of PAH cases would be misdiagnosed. Pyramid charts for both scores are shown in figure 1. Receiver operator curve analysis demonstrated an area under the curve of 0.82 for the OPTICS score and 0.85 for the H2FPEF score.

Conclusion This study further demonstrates the OPTICS scores ability to non-invasively detect between 1 in 3 and 1 in 4 cases of postcapillary pulmonary hypertension whilst maintaining a low false positive rate. The H2FPEF score had a greater sensitivity, yet crucially a lower specificity and hence a higher risk of misdiagnosing true PAH.

SELEXIPAG TITRATION AND DOSING PATTERNS IN PATIENTS WITH PULMONARY ARTERIAL HYPERTENSION (PAH) IN A REAL-WORLD CLINICAL SETTING: INSIGHTS FROM THE EXPOSURE STUDY

1-5MK Johnson, 7TI Lange, 6Sädeberg, 8Biederlamm, 4A Müller, 9X Luik, 7P Escribano, 7S Gaine. 2Scottish Pulmonary Vascular Unit, Golden Jubilee National Hospital, Glasgow, UK; 1University Medical Center Regensburg, Regensburg, Germany; 3Department of Public Health and Clinical Medicine, Cardiology and Heart Centre, Umeå University, Umeå, Sweden; 4Actelion Pharmaceuticals Ltd, Allschwil, Switzerland; 5StatFinn Estonia OÜ, Tartu, Estonia; 6Pulmonary Hypertension Unit, Cardiology Department, CBERCV, Hospital 12 de Octubre, Madrid, Spain; 7National Pulmonary Hypertension Unit, Mater Misericordiae University Hospital, Dublin, Ireland; 8Queen Elizabeth University Hospital and Gartnavel General Hospital, Glasgow, UK.

Selexipag is an oral IP prostacyclin receptor agonist approved for the long-term treatment of pulmonary arterial hypertension (PAH) in adults with WHO FC II/III symptoms. Selexipag is administered twice daily (b.i.d) and titrated to the patient’s highest tolerated dose. In the GRIPHON trial, treating patients with an individualized dose, identified during a 12-