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

S78 PREDICTING POSTCAPILLARY PULMONARY HYPERTENSION: VALIDATION OF THE H2FPEF AND OPTICS SCORES

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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.

S79 SELEXIPAG TITRATION AND DOSING PATTERNS IN PATIENTS WITH PULMONARY ARTERIAL HYPERTENSION (PAH) IN A REAL-WORLD CLINICAL SETTING: INSIGHTS FROM THE EXPOSURE STUDY

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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-

Abstract S78 Figure 1

week titration period, provided a consistent beneficial outcome across doses. Here we describe the titration and dosing of selexipag in a real-world clinical setting, using EXPOSURE study data.

EXPOSURE (EUPAS19085) is an ongoing, international, multicentre, prospective, observational study of PAH patients initiating any new PAH-specific therapy. These analyses include all patients receiving selexipag with follow-up data. Titration was considered complete once a stable dose was taken for ≥ 3 weeks after the highest identified dose. Patients entered the maintenance phase on week 2 of that stable dose. Titration and maintenance phases were defined after data collection. Patient characteristics and treatment patterns at selexipag initiation are described. Values are median (range) unless otherwise stated.

As of June 2020, 300 selexipag-treated patients had follow-up data. At selexipag initiation: age and time from PAH diagnosis was 60 (18–87) and 2.7 (0.0–51.3) years; 70% patients were female and 60% were at intermediate risk of 1-year mortality (COMPERA approach). PAH was classified as idiopathic (51.3%), associated (41.7%), drug/toxin induced (2.7%), and heritable (4.3%). Exposure to selexipag was 5.4 (0.2–29.0) months. Most (77%) patients initiated selexipag as part of a triple combination therapy regimen. At last available information, the titration was completed in 224 (75%) patients, undergoing in 55 (18%) and not completed in 21

(7%). Titration duration was 7.3 (1.0–25.7) weeks. The first maintenance dose achieved was 800 (50–2000) μg b.i.d and lasted for 15.7 (2.0–118.3) weeks. During the selexipag exposure period, 168 (75%) patients remained on the same maintenance dose, 41 (18%) patients had one dose adjustment and 15 (7%) patients had ≥ 2 dose adjustments.

With a median duration of 7.3 weeks, the observed titration period is in line with GRIPHON and the selexipag prescribing information. Approximately 25% of patients had further dose adjustments following titration.

Please refer to page A189 for declarations of interest related to this abstract.

S80

THE DIAGNOSIS AND MANAGEMENT OF CATHETER-ASSOCIATED UPPER-EXTREMITY DEEP VENOUS THROMBOSIS (CA-UEDVT): A SYSTEMATIC REVIEW

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Introduction and Objectives Central venous catheters are a main risk factor for deep venous thrombosis of the upper extremity (UEDVT), with 93% of all UEDVTs being catheter-

PRISMA 2009 Flow Diagram

Abstract S80 Figure 1