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Chest Wall International Group (CWIG)

The importance of the diagnosis of the malignant tumour of the myelin sheath associated with neurofibromatosis: Case report

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Introduction. Malignant neoplasms of the peripheral nerve sheath (TMBNP), also called malignant Schwannoma, are relatively rare neurogenic neoplasms. They are usually found in the mediastinum, retroperitoneum and, rarely, in the thoracic wall. In addition, it has an intimate relationship with neurofibromatosis type 1 (NF1). We report the case of a 17-year-old male patient with multiple thoracic wall nodules whose immunohistochemical result was compatible with NMBBP. A review of the literature on TMBNP and NF1 was carried out, in addition to a case report of a patient with such neoplasia submitted to lesion resection.

Case. A 17-year-old male arrived at the service with complaint of chest pain left 5 months ago. Patient with NF1. Magnetic resonance imaging of the chest revealed multiple nodular formations in the thoracic wall, bilaterally compatible with neurofibromas and a larger lesion of about 2 cm, leading to erosion of the 7th costal arch to the left, with pathological fracture. The patient was sent for a biopsy with the removal of the 7th left costal arch and sent to the anatomopathological study, which showed malignant neoplasia composed of spindle cell fascicles with hypercellular areas, cellular pleomorphism and atypical mitoses. An immunohistochemical study showed focal expression of protein S-100 and collagen IV, in addition to reduction of the expression of H3.3K27. These results are compatible with TMBNP. The patient subsequently underwent a thoracotomy for resection of lesion with broad margins, in addition to reconstruction of the chest wall using Bio-A screen.

Conclusion. The reported case is a relevant demonstration of this rare disease and its description in the literature, being important to emphasise the early diagnosis and the curative surgical approach in this patient. In addition, this differential diagnosis is considered in NF1 patients in order to increase the survival of these patients with early diagnosis, in addition to reducing the morbimortality associated with this disease.

Surgical correction of Poland syndrome 45 days old infant with an autologous graft of the costal arch and sternum and muscular flaps at a teaching hospital

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Introduction. Poland's syndrome is a rare genetic disease characterised by absence of the pectoralis major muscle and multiple anatomical variations. The present study reports the surgical correction of this syndrome in an infant of 45 days of life, with the aim of demonstrating the possibility of early correction.

A review of the literature on Poland syndrome and chest wall reconstruction was carried out, as well as a case report with review of the medical record for 1-year follow-up of infants.

Case. A 45-day-old infant with moderate respiratory discomfort, tachypnoea, motility, difficulty in breastfeeding with frequent choking, and chest asymmetry with left depression. A chest X-ray was performed, which revealed dextrocardia and asymmetry in the left costal arches. Computed tomography with 3D reconstruction of costal arches showed poor formation of lower costal arches to the left, without sternal cover, and agenesis of the pectoralis major muscle, absence of breast and adipose tissue on the left, and discrete scoliosis. Therefore, as the patient developed respiratory discomfort and dysphagia, the surgical approach was programmed for the 45th day of life.

During surgery, a sternal medial incision was performed, exposing the flap where there was absence of the major pectoralis muscle and failure to close the seventh, eighth and ninth left costal arches. A perichondrium graft for correction of the arches was performed with reconstruction of the thoracic wall with the rotation of the large dorsal muscle.

Conclusion. The treatment of this syndrome is most often carried out in a later age group, in adults and adolescents. However, in this case, the patient evolved from birth with respiratory distress, in addition to dysphagia and insufficient weight gain. Therefore, surgery was proposed for the patient. In the immediate postoperative period, the patient evolved with an important improvement in the respiratory pattern; and late with growth normalisation, confirming that the surgical process in this infant was very important to ensure healthier development.

Desmoid tumour in a male patient without risk factors with thoracic wall reconstruction using omentum cover: Case report

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Introduction. Desmoid tumour - or aggressive fibromatosis - is a rare benign neoplasm of origin in the musculo-aponeurotic tissues, with low metastatic content and high local infiltrative potential. It accounts for about 3% of soft tissue tumours and the highest discrete prevalence among females of childbearing age, usually in individuals between 15 and 60 years of age. The objective of this study is to report a case

of uncommon benign neoplasia in a young patient with no risk factor, whose treatment was essentially surgical with complete resection of the lesion and good clinical response to reconstruction of the chest wall using the omentum flap. A review of the literature on desmoid tumours and reconstruction of the thoracic wall was carried out, besides a case report of a patient with such neoplasia who underwent resection of the lesion.

Case. A 39-year-old male without comorbidities, with a history of chest pain for 1.5 years and a tumour in the right hemithorax. The patient had no history of trauma or previous surgeries. Thoracic tomography showed an oval, expansive, soft tissue attenuation located in the muscular plane of the thoracic wall, in the antero-inferior region to the right at the thoraco-abdominal transition, measuring 6.0 × 2.4 × 5.0 cm. A biopsy of the lesion had anatomopathological and immunohistochemical results compatible with desmoid fibromatosis. The surgery was performed with the resection of a piece of 15 × 12 × 3.5 cm, with a mean margin of 2 cm with the removal of the parietal pleura and 6 costal arch segments, as well as the reconstruction of the chest wall with the placement of a Marlex screen, sternal prosthesis of methacrylate and rotation of omentum, and complemented with cutaneous tissue graft.

Conclusion. Desmoid tumours are uncommon, locally aggressive, benign tumours, with high local recurrence, even after adequate treatment. Patients with extensive thoracic neoplastic lesions become a challenge for thoracic wall reconstruction. We present this case to highlight the importance of this differential diagnosis among the thoracic neoplasias and to emphasise that the surgical treatment must have detailed planning in order to avoid complications for the patient.

Chronic pain after Nuss bar procedures for pectus excavatum can be caused by low grade infection

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Introduction. The Nuss bar procedure is nowadays a welcome and relatively safe operation to correct the pectus excavatum (PE). The chronic infection rate reported in the literature is low, between 1 and 3%. In our 8-year experience, we noted some patients with chronic pain after the Nuss procedure, who happened to have low-grade infection.

Objectives. To analyse the patients and their treatment and outcome.

Methods. We studied all charts from our series of Nuss bars for PE, performed since 2011. The patients who were diagnosed with chronic infection, or low-grade infection, were selected. We studied their symptoms, blood test results, microbiology test results, reoperations, the antibiotics they had and the clinical outcome.

Results. In our series of 240 patients who underwent Nuss bar surgery for PE since 2011, we found 6 patients (2.5%) who had chronic infection. They all presented with chronic pain, and limited or no redness of the skin. The blood tests were abnormal in 5 of

the 6, and the microbiology cultures showed bacteria in 3 of the 6 (*Cutibacterium* (n=2) and *Staphylococcus* (n=1)). Three patients had their bars removed, and two had local drainage procedures. Five patients received antibiotics for weeks (clindamycin and/or rifampicin). One patient had no need for antibiotics after bar removal. The chronic pain was relieved by the surgery and /or antibiotics in all 6 patients.

Conclusion. Chronic pain after Nuss bar procedure for PE can be caused by chronic (low-grade) infection. In our series, this was found in 2.5% of the patients, in concordance with the literature. Removal of the bar(s) and/or antibiotic treatment for weeks relieved the pain in our patients.

Pulmonary functional assessment 5 years after chest wall reconstruction with methyl methacrylate for oncologic resections

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Introduction. It is uncertain whether rigid chest wall reconstruction after chest wall oncologic resections would result in limited pulmonary reservoir.

Objective. We sought to determine the long-term impact of methyl methacrylate chest wall reconstruction on pulmonary function test (PFT).

Methods. A retrospective observational study was performed of all adult patients with chest wall malignancy who received reconstruction with methyl methacrylate between 2007 and 2014. Lung cancer patients were excluded. Standard laboratory-based PFT was performed preoperatively and at the 5-year follow-up. The primary outcome was the changes in forced vital capacity (FVC), forced expiratory volume in 1 second (FEV₁), and total lung capacity (TLC). Secondary outcomes included prosthetic complications, surgical mortality, and time to return to work

Results. A total of 19 patients met the study inclusion criteria; 11 patients were males, with a mean age of 58 years, and 9 patients had a history of smoking. No preoperative respiratory comorbidities were noted. Resection was indicated for primary chest wall malignancy in 10 patients and for secondary chest wall malignancy in 9 patients. Anterolateral resections were performed in 16 patients and sternal resections in 3 patients (1 full sternectomy, 2 partial sternectomy). The mean number of ribs resected was 4 (range 3 - 8). Preoperatively, PFT did not show any restrictive or obstructive pattern in any patients. At the 5-year follow-up, PFT revealed no significant difference of predicted v. recorded FVC and FEV₁. The median ratio of TLC was shown to be significantly higher than 0.85, indicating prevention of respiratory restriction. No surgical mortality was observed. No patient developed prosthetic complications. All patients returned to work within a mean period of 5 weeks following discharge.

Conclusion. Chest wall reconstruction using methyl methacrylate does not affect pulmonary function or result in limited pulmonary reservoir.

Thoracoscopic cryoanalgesia: A promising strategy for pain control following minimally invasive repair of pectus excavatum

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Introduction. Recent publications report rapid hospital discharge and low requirements of pain control medication after bilateral intercostal nerve cryoablation during minimally invasive repair of pectus excavatum (MIRPE).

Objectives. To report our initial experience with MIRPE.

Methods. We performed a retrospective chart review of patients submitted to intraoperative cryoanalgesia during MIRPE in our institution from September 2018 to March 2019. All patients with pectus excavatum operated on during that period were included. A liquid nitrogen cryosurgery system was used to cool a probe to -70°C . Then, under thoracoscopic control, the probe was directly applied to the intercostal nerve for 2 minutes. This was done bilaterally along five intercostal spaces, from the 3rd to the 7th space. Postoperative pain control was assessed with a Visual Analogue Scale (VAS).

Results. A total of 21 patients were included; 90% were male. The mean age at surgery was 15.7 years (range 12 - 22) and the mean weight was 55.8 kg (range 38 - 73). The mean Haller index was 5.6 (range 3.5 - 16.9), the mean correction index was 41.4% (range 23 - 76.6) and 90% had sternal rotation, 63.2% to the right and 36.8% to the left. Heart displacement was observed 66.7%, all to the left; 66.7% of the patients received 3 bars and 32.3% patients received 2. The mean duration of cryoanalgesia was 60.1 minutes (range 28 - 100). None received epidural anaesthesia. There were no intraoperative complications. The mean length of stay was 51.9 hours (range 28 - 79). Thirty-three percent of the patients did not require opioids for pain control postoperatively, 42.9% required 1 or 2 doses of opioids and 22.8% required more than 3 doses. Analysis of the VAS on postoperative days 1, 2, 7 and 21 were, on average, 3.2, 2.5, 0.9 and 0.4.

Conclusions. Cryoanalgesia during MIRPE allowed a rapid hospital discharge with very good pain control in all cases. Cryoanalgesia has become our first choice of treatment for pain control in thoracoscopic correction of pectus excavatum

Comprehensive analysis of cardiac MRI, pulmonary function tests, and cardiopulmonary exercise tests in 325 pectus excavatum patients: Severity of defect correlates with abnormal cardiopulmonary function

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Introduction. Pectus excavatum (PE) is a common chest wall deformity with variable severity and symptoms. Whether PE has adverse physiological effects or is purely cosmetic remains controversial among healthcare providers and payers. Clear and consistent data that PE compromises cardiopulmonary function remains sparse. We hypothesise correlation between degree of PE and cardiopulmonary dysfunction.

Methods. Results of cardiac MRI (cMRI), pulmonary function tests (PFTs), and cardiopulmonary tests (CPETs) were reviewed in 325 PE patients evaluated at our institution over a 3-year period. Regression modelling was used to determine whether there is a correlation between pectus indices and clinical endpoints of cardiopulmonary function.

Results. Complete data from 325 cMRIs, 268 PFTs, and 256 CPETs were analysed. Eighty percent of the patients were male and 98% were white. The mean (standard deviation (SD)) age was 15.2 (4) years (range 5 - 39); 84% <18 years. The mean (SD) pectus indices were: Haller index (HI) 5.4 (2.3), depression index (DI) 0.63 (0.6), correction index (CI) 33.6% (14.5), and cardiac correction index (CCI) 2.79 (0.93). Measures of cardiac function revealed a mean (SD) right ventricular ejection fraction (RVEF) of 54% (5) and left ventricular ejection fraction (LVEF) of 58% (4), while 14% of patients had RVEF <50%, 22% LVEF <55%, 33% RVEF z-score <-2, and 18% LVEF Z-score <-2. PFTs (FVC, FEV₁, FEV₁/FVC, TLC) and CPETs (VO₂ max, O₂ pulse, work, breathing reserve (BR)) were abnormal in up to 30% of patients. Multivariate analysis revealed significant ($p<0.05$) negative correlations for: 1) HI with RVEF, RVEF z-scores, LVEF, LVEF z-scores, TLC; 2) CI with VO₂ max, O₂ pulse, work; and 3) CCI with FEV₁, FVC. Binary multivariate analysis using RVEF <50%, RVEF z-score <-2, LVE <55%, LVEF z-score <-2, FEV₁<80%, FEV₁/FVC <85%, TLC <80, VO₂ <80%, O₂ pulse <80%, work <80%, and BR <20 revealed significant ($p<0.05$) correlations for: (i) HI with decreased RVEF, LVEF z-scores; (ii) CI with decreased VO₂ max, work; (iii) CCI with decreased FEV₁, FVC, BR; (iv) DI with decreased TLC; (v) age with decreased RVEF z-scores, LVEF, LVEF z-scores, FEV₁/FVC; (vi) dyspnoea with decreased RVEF. Female gender was favourable for improved PFTs and CPETs ($p<0.05$).

Conclusion. Contrary to popular thought, PE is not just a cosmetic issue. It is associated with heart and lung dysfunction. Both RV and LV function are affected. The severity of pectus defects correlates with dysfunction, which is also influenced by age and gender. HI, CI, and CCI may be the most useful predictors of impairment.

Experience in diagnosis and treatment of 53 cases of primitive neuroectodermal tumour of the chest in children

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Introduction. Primitive neuroectodermal tumour (PNET) is a rare and highly malignant tumour of the chest. Tumour biopsy can be one of the effective methods to establish the diagnosis. The treatment of PNET requires multiple therapies.

Objective. To review and analyse the experience of clinical characteristics, pathological characteristics, diagnosis and treatment of chest PNET in children.

Methods. We retrospectively reviewed the data of 53 patients with chest PNET diagnosed by surgery or biopsy from September 2007 to March 2019 who were enrolled in our study.

Results. The male ($n=31$) to female ($n=22$) ratio was 1.4:1. The common manifestations were the progressive enlargement of chest wall mass, fever, chest pain or shortness of breath. The mean age was 8.6 years and 16 cases were diagnosed by fine-needle aspiration or surgical biopsy. A total of 43 patients received surgery. The mean blood loss was 50.0 mL, and the mean operation time was 106.5 min. The tumour size ranged from 2.5 - 16.0 cm in greatest dimension. The immunophenotype of CD99 and Ki-67 were positive in most of the cases.

Conclusion. PNET is a rare and highly malignant tumour in the chest. Tumour biopsy can be one of the effective methods to establish the diagnosis. CD99 and Ki-67 have important reference value for the differential diagnosis of PNET. The treatment of PNET required multiple therapies.

Innovation in adult pectus repair D2R (Deconstruct to Reconstruct)

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Introduction. New technique in the repair of pectus excavatum in the adult population. Pectus excavatum is the most common chest wall abnormality in children affecting approximately one in 400 births. The defect is marked by three constant and variable defects: (i) depression of the sternum inward, distal to the sternomanubrial angle, (ii) malrotation of the sternum, usually to the right of the chest, and (iii) crowding of the costochondral plates creating the point of inclination. These deformities can cause profound physiological disturbance which may become symptomatic as the patients' cardiovascular demands increase beyond the point of their function reserve both respiratory and cardiovascular. There is also a component of psychological distress and self-esteem.

Objective. We present an alternative to the correct procedures for correction of adult pectus excavatum.

Methods. The repair in the adult population remains challenging. We are presenting a new surgical technique described as D2R (deconstruct to reconstruct). This new technique involves sternal anterior osteotomy, detachment of costochondral junction from the lower sternum, resection of the xiphoid process, elevation and stabilisation of the sternum with parallel titanium plates, mesh placement and sliding myofascial flap of the rectus abdominus muscles for the closure of the lower thoracoabdominal defect.

Results. We have performed this procedure in 4 patients. The 30 and 90-day mortality were zero. The cosmetic and functional results were excellent. The most common complications were pleural effusion ($n=2$) and incisional hernia ($n=1$).

Conclusion. Although the Nuss bar has been the standard of care in the paediatric population, its application in the adult population has had different success rates.

Modified Nuss procedure v. Ravitch in concurrent repair of pectus excavatum & open heart surgery

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Introduction. Pectus excavatum (PE) is the most frequent congenital anomaly of the chest wall. It can be associated with cardiac disorders need surgical repair. Surgical techniques and timing whether concomitant or staged repairs remain controversial. Here is a case series with either modified Nuss or Ravitch for concurrent surgery and review the literature.

Objective. We present a series of pectus repairs associated with open cardiac procedures, with discussion of the outcomes.

Methods. Seven patients with PE deformity (age range 5 to 26 years) and concurrent heart disease were enrolled. Four patients presented with Marfan's syndrome (2 with mitral valve regurgitation and 2 with aortic root aneurysm) and one patient with Noonan's Syndrome. One patient had previous failed Ravitch with complicated right coronary artery (RCA) to right ventricular outflow tract (RVOT) fistula and one with Pouters chest had ASD and subpulmonic RVOT outflow stenosis. Median sternotomy used for 5 patients with subsequent 2 mitral valve replacement (MVR), 2 Bentall operation plus bullectomy and mechanical pleurodesis in one, one David operation and one repair of RCA fistula with concomitant open Nuss repair in all. In two patients a longitudinal right or left parasternal costochondrectomy was done with MVR in one and closure of ASD and bovine pericardial patch repair of RVOT stenosis in the other with concurrent modified Ravitch technique.

Results. There was no mortality or morbidity in this series. Three patients had staged repair within 24 hours to have safe operations after open heart surgery.

Conclusion. There are few successful reports of simultaneous open heart surgery and pectus repair in adolescence and adult. The techniques of concurrent pectus repair remain a challenging issue with their own pro and cons. Nuss procedure is less invasive than modified Ravitch technique although it is not reported in Pouter's deformity. Ravitch technique is used more in combined surgery due to high familiarity.

Long-term outcome after sternochondral allograft for anterior chest wall reconstruction

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Introduction. Reconstruction of the sternum after wide resection for anterior chest wall tumour and large bone loss after deep sternal wound infection are mandatory. The ideal material for chest wall reconstruction continues to challenge thoracic surgeons.

Objective. To present our long-term outcomes of bone allograft for sternal reconstruction after wide resection.

Methods. Between January 2012 and February 2019, we had 5 cases with sternal tumour including synovial cell sarcoma, myxoid tumour, fibrous tumour, chondroma and recurrent fibromatosis. Five patients with sternal wound infection post cardiac surgery were reconstructed with bone allograft. After full evaluation and obtaining consent for bone allograft, the patients were put on the list for an allograft harvesting from a donor or post-mortem within 24 hours of death. Sternal allograft was used after processing by serial cultures, freezing and sterilising with ethylene oxide. Allografts were fixed with titanium microfixation with locking screws in 6 and without locking screws in 4. Thoracic soft tissue reconstructed with bilateral pectoralis major muscle flap in 9 with added omental flap in one and bilateral latissimus dorsi in the last patient who had recurrent fibromatosis and 2 previous operation, as well as radiotherapy.

Results. The operations were uneventful. CT scan 6 months to 6 years after operation were reported normal with some area of vacuolisation and decreased mineralisation. Four patients had partial or total allograft dislodgement. Two patients had reoperation to revised allograft fixation within 9 months to 6 years. The first patient (BMI = 40) developed deep soft tissue and breast infection. She managed with water jet hydrotherapy and negative pressure wound therapy. The other patient with diabetes mellitus and post coronary artery bypass graft (CABG) surgery had infection who needs partial resection of lower sternal allograft with revision of soft reconstruction. In the patient with recurrent fibromatosis and radiotherapy, skin flap necrosis occurred and she needed two revisions of the skin flaps. Two obese patients with post CABG and large bone allograft died 35 to 45 days postoperatively due to cardiac events unrelated to chest wall reconstructed.

Conclusion. This technique has a new era in anterior chest wall reconstruction which provides good functional and cosmetic results. Allograft procurement from a donor and technique of bone fixation to prevent dislodgement are major issues to be considered.

STRATOS system failure in adolescent carinatum surgery

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Objective. To evaluate our experience with the Strasbourg Thoracic Osteosyntheses System (STRATOS) for repair of pectus carinatum for outcomes, complications, and risk.

Methods. Retrospective interrogation of a chest wall database and case notes of patients who underwent surgical correction of Pectus Carinatum using the STRATOS system.

Results. The STRATOS system was used for sternal stabilisation in 5 male patients who underwent a modified Ravitch operation by one surgeon. Age range was 14 - 17 years (mean 15.8). Four defects were classed as 'severe', while one was 'moderate/severe'. Four were symmetrical. Three failed dynamic bracing before surgery.

Two bridges were used per patient. Hospital stay was 5 - 7 days (mean 6). There were no early postoperative complications. Cosmetic appearance was classified as 'excellent' in four and 'good' in one. All patients were satisfied and discharged from follow-up.

Two patients were re-referred at 898 and 1 154 days after repair, reporting newly prominent metalwork. One described discomfort. Chest radiographs confirmed mechanical failure of the STRATOS system in both. The remaining three patients were invited for review. Radiographs confirmed failure of the STRATOS system in all.

All patients had surgery to remove the fractured/displaced metalwork. Four underwent removal of central components to the level of the hinges. One patient later requested removal of rib clips. The remaining patient had one procedure to remove all components.

Conclusion. In this small series, both bridges failed in each patient. The shortest time to detecting failure was 26 months. At the time of using the STRATOS system, removal was optional. The manufacturer now recommends removal after 2 - 3 years. In these cases, all metalwork was anterior to the thoracic cage so failure was less likely to cause serious injury. Recommended use of the STRATOS system retrosternally for pectus excavatum risks serious consequences in the event of fracture or failure. We no longer use or recommend this system for correction of pectus deformities.

Magnetic resonance imaging for pectus excavatum assessment in children

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Introduction. Efforts for radiation dose reduction have allowed cardiac magnetic resonance imaging (c-MRI) to play a role in pectus excavatum (PE) assessment. C-MRI provides a precise anatomic study and also a functional assessment of the malformation.

Objective. To describe the findings of c-MRI after we replaced chest computed tomography to c-MRI for the PE preoperative assessment protocol.

Methods. Since mid-2015 to 2018 all patients with severe PE were assessed with inspiratory and expiratory c-MRI. A retrospective analysis of these patients was performed evaluating (i) radiological PE indexes changes during breathing, (ii) cardiac function and (iii) inferior vena cava (IVC) compression compared with healthy controls.

Results. A total of 56 patients met the inclusion criteria. Dynamic imaging showed a significant difference during inspiration and expiration of the Haller's index 3.92 (range: 3.16 - 7.49) v. 5.17 (range 3.91 - 10.9) ($p=0.05$), and correction index (26.92% v. 36.85%, respectively, $p=0.05$). c-MRI analysis disclosed a right ventricle ejection fraction of 50.1% (normal range: 61% (54 - 71%)). Related to IVC, differences were only statistically significant for anteroposterior diameter in males 12.7 ± 0.5 mm (95% CI 11.65 - 13.82 mm) v. 16.5 ± 0.5 mm (95% CI 15.71 - 17.62 mm) ($p=0.000$).

Conclusion. Our results showed that PE assessment by c-MRI allows a radiation-free image of the chest wall deformity during the entire breathing process. Also, it allowed the evaluation of the influence of the malformation on cardiac function and on the IVC. These findings also provide an accurate preoperative planning and surgical evaluation.

Thoracoplasty as a procedure of continuing relevance

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Introduction. Thoracoplasty is operative removal of the skeletal support of a portion of the chest, which involves sub-periosteal removal of a varying number of ribs segment. The objective is to make the unsupported portion of chest wall sink in towards the mediastinum and reduce the size of hemithorax. This partial decostalisation of the thoracic cage is aimed to close persistent pleural space or collapse apical cavities or diseased lung.

Methods. At the National Institute of Tuberculosis and Respiratory Diseases, New Delhi, India, out of a total of 6 443 thoracic surgical procedures done from April 1997 to March 2018, a total of 498 were thoracoplasties and 415 of those were carried out for persistent pleural spaces post-resection or those following empyema management. A total of 62 thoracoplasties were done for massive recurrent haemoptysis in situations where lung resection was not possible and 21 were done to address the problem of persistent sputum-positive state because of drug resistance or otherwise.

Results. Eighteen patients died in the immediate postoperative period as a result of several reasons. There were 62 late deaths on follow-up ranging from 6 months to 10 years. There was significant relief in the symptoms for which the surgery was performed. Twelve patients did not turn sputum-negative despite thoracoplasty in the group taken up for persistent sputum negativity. Only 2 patients in the haemoptysis group required further surgery for continuing haemoptysis. The problem of chest wall sinus was completely addressed in almost all the patients.

Conclusions. Though an old operation, the procedure of thoracoplasty offers significant benefit to patients whenever it is indicated.

Enhanced recovery in thoracic surgery – where do we stand?

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Introduction. The presentation will review the literature around enhanced recovery in thoracic surgery and present the latest research and consensus guidelines. It is aimed at cardiothoracic surgeons and peri-operative physicians caring for patients having thoracic surgery, mainly lung resection.

A rare management of Chilaiditi's syndrome

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Introduction. Chilaiditi's sign is a radiographic term used when the hepatic flexure of the colon is seen interposed between the liver and right hemidiaphragm. In most cases, this interposition is asymptomatic and is often an incidental finding in the elderly. When it is symptomatic, this is Chilaiditi's syndrome.

Case. The authors report a case of Chilaiditi's syndrome in a 56-year-old male patient. Although this is a benign condition with rare surgical indication, it has great importance for implying surgical emergency in cases where such condition is equivocally diagnosed as pneumoperitoneum. In our case, colonic resection was decided on elective basis as a trial to improve the patient's respiratory symptoms which was resistant to conservative lines of treatment. A literature review is performed with emphasis on pathophysiology, diagnostic work-up and treatment of this entity.

Conclusion. Although Chilaiditi's sign is a rare entity overall, this challenging diagnosis should be considered when a patient has a radiographic finding of air below the right hemidiaphragm and presents with abdominal and / or respiratory symptoms. Our patient illustrates a rare but clinically important entity as a combination of chronic respiratory difficulty and chronic abdominal distension with aerophagia. This study is a trial to have a solution for Chilaiditi's syndrome associated with clinical and radiological evidence of respiratory distress that is resistant to conservative lines of management. Hopefully, in the future, more research work may prove or disprove elective surgery to sort this problem out. Attention should be paid to factors predisposing to the development of Chilaiditi's syndrome and treatment should be altered accordingly.

Unexpected finding during minimally invasive repair of pectus excavatum

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Introduction. Most patients undergoing minimally invasive repair of pectus excavatum (MIRPE) do not have any other diseases than the thoracic deformity, resulting in an uneventful thoracoscopy. We experienced massive intrathoracic adhesions in one patient during MIRPE unexpectedly.

Case. A 16-year-old boy presenting with symmetric pectus excavatum was scheduled for MIRPE. For the patient's history, only bronchial asthma was reported by the mother. Preoperative diagnostics, including chest X-ray, echocardiography, EKG and lung function test, showed unsuspecting findings. Starting with thoracoscopy, many adhesions in the right thoracic cavity occurred. Some adhesions were detached and the lung itself appeared normal. Due to the unexpected adhesions with unknown underlying cause the planned MIRPE was canceled. After identifying the underlying cause of the adhesions postoperatively, MIRPE including release of the adhesions was planned a few days later.

Conclusion. We present a case in which MIRPE was canceled due to unexpected adhesions of the right lung. The underlying cause could not be seen in the preoperative diagnostics.

Minimally invasive repair of pectus excavatum (MIRPE) without happy ending: Analysis of severe complications related to MIRPE (excluding cardiac injuries)

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Introduction. This review serves to highlight the dangers of a simple operation.

Objective. To warn young surgeons of the potential dangers.

Methods. Review of the literature and personal communications.

Conclusion. Despite its minimally invasive approach, with the widespread use of the minimally invasive repair of pectus excavatum (MIRPE) procedure the character and number of complications have increased. Many studies report near-fatal complications, not only during bar placement, but also during bar removal. Of course, cardiac injuries and/or lesions of the mammary artery represent the most common complications. However, an increasing number of studies report on rare but severe complications excluding cardiac lesions. The results of a selective review of the current English language literature with focus on these rare complications will be presented.

Suction cup treatment for pectus excavatum

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Introduction. Non-surgical measures, such as vacuum bell therapy, were established as useful complements to treat PE patients ~15 - 20 years ago.

Objectives. Review and personal series.

Results. Within the last 5 - 10 years, an increasing number of studies were identified reporting on conservative treatment of PE using the vacuum bell. There were no randomised and/or prospective studies comparing conservative treatment v. surgical repair or conservative treatment v. no specific therapy. Variables predictive of an excellent outcome could be identified. Especially in younger PE patients, conservative treatment is reported with increasing frequency.

Conclusion. The choice of treatment methods is mainly dependent on patient's age at diagnosis and severity of the PE. However, specific treatment is not necessary in every PE patient, but follow-up is important, especially in paediatric and adolescent patients. Non-surgical treatment seems to represent the first step of specific therapy in the majority of PE patients. Patients and parents may appreciate if the surgeon offers all treatment modalities.

Thoracoscopic assisted fixation of rib fractures and lung repair in traumatic chest wall injury

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Introduction. Surgical fixation of multiple level rib fractures requires long and multiple thoracic incisions to expose multiple levels of fractured ribs. However, by thoracoscopic assisted, we performed multiple rib fixation using 4-cm length incision and additional camera port in the case report.

Case. A 58-year-old male with left 4th, 5th, 6th and 7th rib fractures by blunt trauma was referred to our hospital for surgical correction of the herniated lung into fractured rib segments and continuous air leak through chest drain. We planned the reduction of herniated lung and displaced rib, and thoracoscopic rib fixation using multi-hole titanium plates and a chest drain site.

Results. Through a 4 cm incision and one 5 mm camera port, the herniated lung was corrected and pushed back to the chest cavity after careful dissection of fractured ribs. We repaired the lacerated lung parenchyma through the port. The fractured ribs were repositioned and fixed with multi-hole titanium plates. For the fixation of the 4th rib, impossible to screw through the incision, we made an additional 5-mm incision at anterior chest and successfully fixed the high-level rib fracture guided by a thoracoscope. The wound retractor at the muscle layer often helped minimise the wound during the procedure and thoracoscope. The patient was discharged after 5 postoperative days.

Conclusion. Multiple level rib fixation guided by thoracoscope is a safe and feasible option for reducing wound and postoperative morbidity.

The cross-bar technique: Paving the road to remodelling of the entire chest wall

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Introduction. Pectus excavatum repair with pectus bars has been advancing. The cross-bar technique is designed to position the bar accurately and to cover a wider area of the chest wall.

Objective. To prove the efficacy and safety of the cross-bar technique.

Methods. As an important technique for our 'Entire Chest Wall Remodelling' approach, we placed two bars crossing on the target since 2016. The technique is unique in that the lifting forces of the two bars converge on the single target. The second bar attacks at the same spot for the further elevation of the resistant and inflexible chest wall. Additionally, the cross-bars cover more area to the lateral chest wall even lower than the xiphoid. The indications and complications of the cross-bar (group C) and parallel bar (group P) techniques are compared.

Results. From January 2016 to March 2019, 274 patients underwent multiple bar pectus excavatum repair using the bridge technique for bar fixation: 168 cross-bar and 106 parallel-bar cases. The indications

for the cross-bar technique included acute xiphoid depressions, inflexible focal chest wall depression, bilateral costal depressions lower than the sternum, reoperation cases difficult to locate the bar on target, and long deformities with Grand Canyon type. In group C, 51 (30%) received an additional uppermost bar placement (triple bars in 'XI' fashion) to cover the upper chest wall. The median age was 16 years (range 4 - 42). There was no bar displacement case in both groups. Postoperative complications occurred in 14 patients (5%): 9 in group C and 5 in group P. Complications included thoracic outlet syndrome ($n=3$), pneumothorax ($n=9$), and pleural effusion ($n=2$). Late complications in group C were wound infection ($n=6$) and pleural effusion ($n=2$). The overall complication rate in the cross-bar group was not different from the parallel-bar group (11% v. 13%; $p=1.00$).

Conclusion. The cross-bar technique is a unique, inspiring technique that makes all the momentum forces converge into a single spot and covers a wider area of the chest wall without adding risk. Combining the cross-bar technique and the bridge fixation seems to be the best way toward the risk-free remodelling of the entire chest wall.

Surgical repair of pectus carinatum and mixed deformities: The sandwich technique and magic string procedure

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Introduction. The sandwich technique (SWT) is a novel technique that enables the repair of pectus carinatum and pectus excavatum-carinatum complex (PCE mixed deformity) by means of press-molding the deformed chest wall between two bars. Whereas, the technique using only external bar compression has been often incomplete in asymmetric or mixed deformities.

Objective. To verify the surgical indication, elaborate on the details of the technique; and appraise the early results.

Methods. Since we first started in January 2007, various forms of pectus carinatum and pectus excavatum/carinatum complex were treated with SWT. Until March 2019, 324 consecutive cases of SWT and the magic string technique were analysed retrospectively. The sandwich techniques using the internal and external pectus bars were for the repair of pectus carinatum. For the concealed carinatum component, which became prominent after PE repair, the magic string technique has been applied since 2015, where the external thick string (instead of the pectus bar) was used to compress the protuberances after the excavatum repair with the internal pectus bar placement.

Results. The indication for the sandwich repair with the external and internal bars was 65 pectus carinatum varieties: 10 pectus carinatum per se, 25 pectus excavatum/carinatum complex, and 30 emerging concealed carinatum after excavatum repair. The 235 patients with concealed carinatum component of pectus excavatum, often seen in the Grand Canyon type, were managed with the magic string technique. Early postoperative complications occurred in 5 (1.54%) patients; i.e. pneumonia ($n=2$) and pneumothorax ($n=3$). Overall, SWT achieved near-complete resolution of carinatum component. The median asymmetry index was 1.1 (1.0 - 1.28) preoperatively and 1.0 (1.0 - 1.06) postoperatively ($p<0.001$). The magic string

procedure also accomplished almost symmetric configuration without significant residual carinatum components. The median asymmetry index was 1 (1.0 - 1.22) preoperatively and 1 (1.0 - 1.1) postoperatively ($p<0.001$).

Conclusion. The sandwich technique that press-molds the deformed chest wall between the external and internal bars seems effective in treating pectus carinatum. This technique was successfully expanded to the PCE mixed deformity, achieving post-repair symmetry. The magic string technique uniquely offers cosmetic perfection by eliminating concealed mild focal protuberances combined with pectus excavatum, especially in the Grand Canyon deformity.

Elusive ideal pectus repair at Dr George Mukhari Academic Hospital, Pretoria South Africa: A case series

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Introduction. Among chest wall deformities, pectus excavatum is the most common (90%), followed by pectus carinatum (5 - 7%), cleft sternum, pentalogy of Cantrell, asphyxiating thoracic dystrophy, and spondylothoracic dysplasia. An estimated 1 in 300-400 births is associated with pectus excavatum with a male-to-female ratio of 3:1. 90% of cases are diagnosed within the first year of life most of which are typically noticed at birth. During rapid bone growth in the early teenage years there is worsening of the pectus deformity appearance and the onset of symptoms are usually reported.

Cases. A series of 4 patients with pectus deformities (3 pectus excavatum and 1 pectus carinatum) are reported that presented in Dr George Mukhari Academic Hospital in period of 1 year. 3 patients were treated with a modified Ravitch procedure (2 pectus excavatum and 1 pectus carinatum) and 1 (pectus excavatum) was initially treated with a Nuss procedure 2 years prior, but subsequently received a modified Ravitch repair. At our institution we usually see ~1 patient per year but it has subsequently increased to 4 patients in 2018.

Conclusion. The ideal treatment for pectus excavatum has not been identified. However, clinical and radiological parameters assist with the morphology and classification of pectus excavatum which assist with deciding on which management approach should be used, though sources advocate different methods supported by good outcomes according to their unit expertise.

Foreign body aspiration: Still a reality

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Introduction. The entity of foreign body aspiration is very real and as thoracic surgeons we are faced with this condition numerous times during our training and our careers. Many factors can play a role in children especially the need to explore may also lead to inadvertently placing new or shiny objects into their mouths and subsequently inhaling them. Whether by misadventure or accident, or even just uncoordinated movement when eating, foreign body aspiration (hereafter FBA) continues to occur and presents itself often as an

emergency requiring urgent intervention as the patient suffers from a compromised airway.

Methods. My proposal is that I would offer up a presentation on FBA; with a focus on the South African landscape, while looking at the access patients may or may not have to a skilled surgeon to help them in their time of need. Some recent examples from the news may be quoted (including tragic stories of death to those who do not make it to a facility with a competent surgeon in time). The literature will also be searched in order to compare, relate to, and learn from the experiences of others in our field on a broader spectrum.

I would also like to open up a discussion regarding the training of even more surgeons (perhaps our general surgery colleagues who vastly outnumber us) as to being skilled in the manner of managing cases of foreign body aspiration as thoracic surgeons are often few and far between and patients may need to be transported many hours away to get the help they need. An invitation to the audience during the presentation would invite the surgeons present to share some of their own stories. We all have these stories of the threatened and having that limited window wherein we need to get the job done.

I would look at the methods available for managing FBA, the mainstay in most government hospitals still being rigid bronchoscopy. But also to have a look at the instrumentation and use of flexible bronchoscopy.

Results. What if bronchoscopy should fail? In that conundrum, what are the surgeon's options in order to provide the best outcome for his patient? I believe that this has the potential to be an interesting presentation and refreshing to take another look at something thoracic surgeons may come to find commonplace.

Cross-bar for Nuss procedure – better correction but more complications

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Objective. Report of advantages and disadvantages of new technique of Nuss procedure.

Methods. We conducted a retrospective analysis of 231 pectus excavatum cases operations between 2016 and 2018. Since November 2016 in difficult for correction deformities (local deep deformity, significant rigidity of chest wall, costal flaring) cross-bar technique became one of our methods of pectus excavatum correction. It was totally applied in 57 cases. Bars were implanted usually first right to left and up to down direction. After visual evaluation of result, additional bar was implanted left to right and up to down. For deep and rigid chest wall 3 bars were implanted - cross-bar plus 1 horizontal bar usually at the 3rd intercostal space.

Results. The aesthetic result of cross-bar correction was subjectively better (assessment intraoperatively and after surgery) in the majority of cases, however, in 3 cases after trial correction by introducer no added benefit was obtained and correction was accomplished by 1 oblique bar. In cases of deep local and rigid deformity, cross-bar proved to be the best methods of sternal elevation without residual deformity. In cases of simultaneous costal flaring correction, immediate result was also very good.

There were no differences in pain and time of rehabilitation in cross-bar compared with regular cases. During follow-up there was one case of left-to-right dislocation of one bar, and one case of 45-degree rotation of one bar. There was no need for revisions. In 12 cases, clinically significant amounts of pleural effusion resulted in the need for thoracocentesis and in 4 cases, readmission for treatment. In one paediatric case, after initial success, significant carinatum overcorrection developed.

Conclusion. Cross-bar correction of pectus excavatum extends the possibilities of Nuss procedure; however, at the price of an increased complication rate. It should be used only when it is strictly indicated.

Female pectus excavatum patients – gestation and delivery with pectus bar and before correction

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Objective. Presence of severe pectus excavatum (PE) as well as condition after correction but with bar in place is not well known if it will not harm gestation and delivery.

Methods. During the period of 2004 - 2018, 9 females aged 23 - 41 had history of gestation and delivery. All patients were interviewed retrospectively using a specifically designed questionnaire.

Results. The period of gestation was uneventful in both groups. All women in group 1 completed pregnancy successfully with natural childbirth. In the group with a bar in place, women got pregnant 1 month ($n=2$), 1 year ($n=2$), 2 years ($n=1$), and 3 years ($n=1$) after correction. None of the women noted any additional complaints in the chest area during pregnancy. There were 2 cases of caesarean section and 4 cases of natural childbirth. All the women were warned by gynaecologists about possible complications; however, in all cases the warning was dictated only by opinion and not supported by previous experience of gynaecologist. During the period before childbirth all women considered delivery by caesarean section, however it was done only in 2 cases. The rest of women were against caesarean section. One woman could not get pregnant for 3 years but was pregnant 1 month after correction.

Conclusion. Women can successfully complete pregnancy both before correction and with bar in place. Common warnings of gynaecologists about possible complications are probably due to fear of unknown condition. Further data collection will clarify whether there are any contraindications for gestation and delivery with bar/bars in place

The application of three-dimensional body surface scanning in the management of pectus carinatum

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Introduction. Pectus carinatum (PC) is considered a treatable chest wall deformity, especially with increased use of non-invasive, compressive

brace therapy recently. However, to date, imaging methods, such as chest computed tomography (CT) and chest X-ray, which unavoidably exposes patients to radiation, are mostly used to measure the shape and degree of correction.

Objective. We investigated the efficacy of a radiation-free method, i.e. three-dimensional (3D) body surface scanning, and compared it with chest CT.

Methods. This prospective observational study included 64 PC patients who underwent compressive brace therapy at our institution between July 2017 and February 2019. All patients wore the brace for at least 20 hours per day for two weeks and 12 hours per day for 6 months, and they underwent chest PA, lateral chest CT and 3D body surface scanning. We compared all these methods.

Results. The average (standard deviation (SD)) age was 10.52 (4.21) years; and 56 patients were males. The Haller index (SD) was significantly increased from 2.22 (0.26) to 2.69 (0.50) after the treatment ($p=0.001$). The mean (SD) protrusion difference was 21.06 (18.42) mm on 3D body surface scanning, while it was 13.44 (9.39) on chest CT. There was no significant difference between the two modalities. Moreover, there was a significant correlation between the two modalities ($p=0.001$; $R^2 = 0.40$).

Conclusion. According to the findings of this study, 3D body surface scanning seems to be an effective, radiation-free alternative to chest CT.

Influence of sternoclavicular joints on computer simulation models of patient-specific Nuss procedure in the pectus excavatum using finite element analysis

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Introduction. The Nuss procedure which is a surgical treatment for Pectus Excavatum (PE) is required an optimised methods for the patient because the surgical outcome is very different depending on the design of the bar, the inserted location, and the degree of sternal elevation. The sternoclavicular (SC) joint is regarded to affect the artificial elevation of the anterior chest wall, there is no model currently considering it.

Objective. We employed finite element analysis, which is a biomechanical method, to analyse the influence of the SC joint.

Methods. Preoperative CT images of PE patient was acquired, and a finite element model of thoracic cage was constructed. After the Nuss bar applied during surgery was inserted into this model, the bar was elevated to 15, 20 and 25 mm in the vertical direction (y -axis) of the coronal plane. In addition, the cases where the influence of the SC joint is removed and the behaviour of the SC joint is constrained in the displacement of the y -axis direction and the rotation of the x - and z -axis were analysed respectively.

Results. The maximum equivalent stress on the sternum, the reaction force by the sternum, and the maximum height of the

elevated chest wall in the y -axis were analysed according to each condition. As the height of the bar was elevated or the constraint of the SC joint was applied, all the physical values increased. In the case of the equivalent stress, the condition was found to increase by 42 – 82 MPa more than the condition.

Conclusion. We concluded that the condition and the condition, which have similarities with the physical behaviour of the chest wall, could not neglect the influence of the SC joint as the difference between the two condition is significant. In addition, since the equivalent stresses on the chest wall were greatly differed according to the elevated height of the bar in condition, the SC joint in the simulation model should be determined to consider their influence in the case of a severe PE patient having a depression from the SC joint level.

Bilateral thoracoscopy for intraoperative intercostal nerve cryoablation during the Nuss procedure: Preliminary experience

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Introduction. Pectus excavatum correction by Nuss technique causes important postoperative pain difficult to control, and long-term pain during the recovery phase. Different strategies have been described for its management. Recently the use of intraoperative intercostal nerve cryoablation for these patients has been published.

Objective. We present our first 2 cases as an alternative pain management for pectus excavatum repair.

Methods. Retrospective observational study of the patients who received a Nuss procedure between March and April 2019. After dissection of a retrosternal window under sternal elevation by crane-technique cryoablation was performed transthoracically using a long 3 mm cryoprobe (Cryo-S Painless). Five intercostal nerves were treated on each side. Analysed variables were: age, type and grade of pectus excavatum deformity, surgical time of cryoablation, postoperative opioid requirements, hospital length of stay, chronic use of pain

Results. In this period of time two patients (males aged 14 and 15 years) underwent the Nuss procedure with cryoanalgesia. The medium Haller index was 4.3 and the correction index was 30%. Medium extra-surgical time for cryoablation was 50 min. In one case, cryoablation was performed before implantation of the Nuss bars, in the second, after. No additional ports or incisions were needed. Both patients received morphine perfusion in the initial postoperative period, which could be retired 48 hours later. Afterwards, pain could be controlled with common analgesics with favourable pain scores and active respiratory physiotherapy. Hospital stay was in both cases 3 days. Oral pain medications could be retired after 12 days in one case and in the other after 25. No complications related to the cryoanalgesia procedure were observed.

Conclusion. Cryoanalgesia seems to be an effective and long-lasting tool for pain management in the pectus excavatum patient. Comparative studies and larger series of patients are needed to prove reduction of hospital stay and pain medication requirements.

Intraoperative cardiologic changes during surgical correction of paediatric pectus excavatum assessed by transoesophageal echocardiography

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Introduction. Cardiac compression in pectus excavatum (PE) is difficult to evaluate by standard preoperative examination. Few studies report cardiac changes by intraoperative transoesophageal echocardiography, and none of them include the paediatric population or the changes in the tricuspid annulus.

Objective. To describe our findings with intraoperative transoesophageal echocardiography during PE correction in paediatric patients.

Methods. We prospectively studied right-heart changes during surgical correction of PE by transoesophageal echocardiography. We measured diastolic diameters of the right ventricle, right atrium and tricuspid annulus, and associated 4D echocardiography to assess the morphology of the tricuspid annulus.

Results. Sixteen patients, with a mean (standard deviation (SD)) age of 13.31 (3.1) years were included between December 2016 and December 2018. The mean (SD) preoperative Haller index was 6.03 (1.5) and the mean (SD) correction index was 46.25% (11.9%). Preoperative transthoracic echocardiography at rest showed mild heart compression in 4 cases. Correction was obtained by the Nuss technique in 15 cases, and Pectus Up in one. Initial transoesophageal echocardiography showed compression of the right heart and deformation of the tricuspid annulus in all cases. During the sternal elevation, diameters of the right atrium, ventricle and tricuspid annulus improved: the mean (SD) augmentation of right ventricle was 5.33 mm (3.48), right atrium 7.25 mm (6.02) and tricuspid annulus 5.41 mm (3.31). These changes were statistically significant ($p=0.05$). Morphology of the tricuspid annulus in 4D was almost normalised.

Conclusion. Preoperative transthoracic echocardiography at rest underestimates right-chamber compression in paediatric patients with PE. Surgical correction improves diameters of the right ventricle, right atrium and tricuspid annulus and almost normalises the morphology of the tricuspid annulus (4D).

Chest wall reconstruction following trauma

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Introduction. The chest wall is the only barrier the chest wall contents have against trauma. The components of the chest wall have an innate ability to withstand trauma but up to a certain extent; the more pliable the chest the better it can withstand trauma. The magnitude of trauma is also important because it also have a bearing on the amount of damage it causes.

Chest wall reconstruction following trauma presents a challenge to thoracic surgeons. Most of the problems arise from the fact that we do

not have clear guidelines or principles to manage such injuries and can only rely on guidelines for chest wall reconstruction for tumour resection. **Case.** A case of a 56-year-old man who presented with an accidental shrapnel injury following the explosion of oxygen canister is presented. The challenges of resultant chest wall injury management are highlighted.

Chylothorax: Still a management dilemma

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Introduction. Chylothorax management remains a challenge and is commonly encountered in the postoperative period by thoracic surgeons. Diagnosis of chylothorax is straightforward; however, its management still poses an enormous challenge and the longer a patient continues to drain the higher the mortality from malnutrition and infections.

Chylothorax is a leakage of chyle from the thoracic duct, aetiology can either be traumatic, non-traumatic, congenital or idiopathic. The most common reason for chylothorax is post-surgery complication during thoracic and cardiac surgery, it is also a problem in patients with malignancies.

Methods. Literature review on the management of chylothorax and challenges thereof. There are no large randomised control trials comparing therapies, therefore the management of chylothorax is still based on clinical experience. The literature review will include the management of low-output v. high-output chylothorax. Postoperative chylothorax and chylothorax due to malignancies, particularly lymphoma, will also be discussed. A review for surgical interventions and various surgical options available, the outcomes and prognosis will be discussed.

Results. Conservative management like pharmacological agents, dietary modifications and other adjuncts is indicated in patients with low-output chylothorax. An urgent surgical intervention is indicated for patients with high-output chylothorax and in patients who fail to respond to medical therapy. Furthermore, the role for intervention radiologists in the management of Chylothorax will be debated, as well as the use of conventional radiation.

Conclusion. The management of chylothorax is challenging, with deleterious effects on patients. Chylothorax still poses a management dilemma. Patients lose chyle, which contains fat, fat-soluble vitamins, proteins, immunoglobulins, lymphocytes which then results in malnutrition and increased risk of sepsis especially in patients with malignancies. Chylothorax management is still dependent on anecdotal experience. Surgical interventions include thoracic duct ligation, chemical or mechanical pleurodesis, pleuroperitoneal shunts, pleurectomy and thoracic duct embolisation and disruption with lymphangiogram.

Haemoptysis secondary to a complicated hydatid cyst of the lung: A case report

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Introduction. Pulmonary hydatid disease is almost exclusively caused by the infestation of the larval stage of *Echinococcus granulosus*. Humans are infected, accidentally, through the faecal-oral route by the ingestion of food and milk, contaminated by dog faeces containing the ova of the parasite, or direct contact with dogs.

Case. We describe an unusual cause of massive haemoptysis in a young male who had bilateral lung hydatid cysts as well as a large splenic hydatid cyst. He underwent bilateral thoracotomies for cyst excision for relief of haemoptysis

Conclusion. This case report suggests a high index of suspicion for pulmonary hydatid cyst as a cause of haemoptysis in a patient who comes from a sheep-rearing community, despite the fact that TB is the most common cause. It also suggests that one should consider operating on the complicated side first, if haemoptysis was the initial presenting complaint of the patient, despite the size of the uncomplicated contralateral site.

Chest wall reconstruction for metastatic tumour

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Introduction. Surgical management of tumours that metastasise to the chest wall is not well defined. Lung tumours that invade the chest wall are T3 according to the 8th edition Lung Cancer Guidelines of the International Association of Lung Cancer. These tumours are considered potentially resectable. Controversy exists regarding the significance of the depth of penetration and whether or not *en bloc* resection of chest wall (v. parietal pleura only) is required for adequate resection. However, when the lung tumour is a secondary metastasis, little is known about the benefit of pulmonary metastasectomy with *en bloc* resection of the adjacent involved chest wall.

Case. Our case is a 16-year-old male patient with previous above-knee amputation for an osteosarcoma. He presented 2 years later with pulmonary metastasis with another metastatic lesion involving two ribs. Bilateral pulmonary metastasectomy was performed and additionally, chest wall resection for of the chest wall metastatic lesion was performed. Chest wall reconstruction with a Gortex Patch and Rib plating was performed.

Conclusion. The patient had an uneventful recovery. However, he presented within 6 months with multiple bony metastatic lesions to the skull, chest wall and lungs.

Small-bore v. large-bore chest drain in the management of patients with pleural effusion at the Lagos University Teaching Hospital, Nigeria

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Introduction. Many researchers have investigated the usefulness of small-bore chest drains in the management of pneumothoraces, pleural effusion and pleurodesis.

Objective. To compare the adequacy of drainage of small-bore catheters against large-bore catheters in the management of patients with pleural effusion.

Methods. Eighty-six patients who presented with pleural effusions were recruited for this study. Two categories of patients were selected randomly and one group was intubated with large-bore (28-32Fr) chest tube, while the other was intubated with small-bore (12-14Fr) chest tube. Adequacy of drainage was compared in terms of post-extubation adequate lung re-expansion, relief of dyspnoea (from NYHA III-IV to I-II), tube blockage, total amount of drainage and duration of drainage.

Results. Comparison between small-bore chest tubes and large-bore chest tubes showed that small chest drains were more desirable to use for the management of pleural effusions. Adequacy of drainage was assessed. In patients with small-bore chest drains, there was adequate lung re-expansion in 31 patients (72.1%), relief of dyspnoea (from NYHA III-IV to I-II) in 30 patients (70%), tube blockage in 5 patients (11.6%) and the mean duration of drainage was 1 - 2 weeks. In those with large-bore chest drains, there was adequate lung re-expansion in 32 patients (74.4%), relief of dyspnoea (from NYHA III-IV to I-II) in 31 patients (72.1%), tube blockage in 4 patients (9.0%) and the mean duration of drainage was 2 weeks. There was no significant statistical difference noted with regards to adequacy of drainage between small-bore and large-bore chest drains. *P*-values were 0.810, 0.815, 0.147, 0.739, for adequate lung re-expansion, relief of dyspnoea (from NYHA grade III-IV to I-II), tube blockage, and duration of drainage, respectively.

Conclusion. Small-bore chest drains were found to be as effective as large-bore chest drains in the drainage of pleural effusions.

Cytology v. biopsy yield in patients with malignant pleural effusion at the Lagos University Teaching Hospital, Nigeria: A prospective review

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Introduction. The yield of cytology and percutaneous pleural biopsies has been investigated by many researchers and the findings have varied.

Objective. To determine the percentage yield of malignant cells from cytology and percutaneous needle pleural biopsies of malignant pleural effusions and to determine the common aetiologies of malignant pleural effusions presenting at the Lagos University Teaching Hospital (LUTH), Lagos, Nigeria.

Methods. A total of 55 consecutive patients with suspected malignant pleural effusion were recruited for this study within a 1-year period. A pleural aspirate was obtained from each patient and cytological analysis was conducted. Percutaneous pleural biopsy was also done for each patient, followed by histological analysis. The diagnostic yields were then obtained and compared for both cytology and percutaneous pleural biopsy. The causes of these effusions were also noted in this study.

Results. The percentage yield of cytology in patients with pleural effusion was 67.3%. Four (7.3%) results were inconclusive. However,

the percentage yield of pleural biopsy was 56.5% (31 patients). The yield was better (81.8%) in patients that had both procedures done with either cytology or pleural biopsy being positive. For both cytology and pleural biopsy, the yield was positive in 40% and negative in 12.7%. When compared with pleural biopsy yield, the cytology yield was not statistically significant ($p=0.252$), however, the yield was higher. In this study, pleural effusion was caused by breast cancer in the majority of the patients ($n=31$; 56.4%). Gynaecological malignancies were responsible for another 25.5%. Together, breast and gynaecological malignancies made up 81.9% of the diagnoses. One patient (1.8%) had oesophageal cancer. Others included chest wall tumour (1.8%) and laryngeal tumour (1.8%).

Conclusion. The yield of cytology in the diagnosis of malignant pleural effusions was higher than that of percutaneous pleural biopsy, though not statistically significant. However, evaluating these patients with percutaneous pleural biopsy as a diagnostic tool increased the chances of arriving at a diagnosis. Thus, percutaneous pleural biopsy is a useful tool in evaluating a patient with pleural effusion. Breast cancer is the leading cause of malignant pleural effusions in our environment.

Compressive brace therapy for pectus carinatum – initial experience

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Introduction. Over the past decade, pectus carinatum orthotic treatment has become the method of choice in many paediatric surgery centres around the world. The development of own compression orthosis at the National Institute of Children's Diseases in Bratislava launched in March 2017. The time from the prototype design to the final version of the brace was ~3 months. A comprehensive orthotic brace treatment of patients with pectus carinatum was thus possible since September 2017.

Objective. To evaluate initial results of compressive brace therapy for pectus carinatum.

Methods. We performed a prospective, single-institution tertiary hospital study to analyse a group of patients using compression orthosis for pectus carinatum treatment between September 2017 and March 2019. Demographic data, anthropometric dimensions and indexes of the chest, data connected with orthosis daily use, patient cooperation evaluation and ongoing treatment outcomes were analysed.

Results. A total 98 patients aged 2 to 19 years with pectus carinatum were examined in our outpatient department over a period of 18 months. Twenty patients met the criteria for inclusion (patients older than 10 years, chondrogladiolar type of deformity, adequate compliance to treatment). Patients with chondromanubrial type of deformity, patients younger than 10 years, patients with insufficient adherence to treatment and those who underwent less than 3 outpatient visits were excluded from the analysis. Regular daily use of the orthosis for about 8 hours was recommended in all patients,

in contrast the largest group of patients carried the orthosis 4 to 6 hours per day (mean 5.58 hours). Significant improvement in chest configuration during first 3 months of orthosis wearing validated by anthropometric examination was noticed in 11 patients (55%). The remaining patients required longer time to first signs of correction.

Conclusion. Conservative orthotic treatment for pectus carinatum patients is more than just a new innovative methods. It does not require hospitalisation, eliminates the risks associated with surgery, anaesthesia and possible postoperative complications. A crucial challenge for the therapeutic team is to ensure adequate patient compliance to achieve the most optimal outcome.

A novel device for sternal closure: Clinical outcomes from a prospective, randomised, first-in-man pilot study

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Introduction. Rigid fixation is the treatment of choice for the management of displaced fractures and osteotomies. However, wire cerclage (WC) remains the predominant closure technique following median sternotomy. WC provides adequate approximation of the sternal halves without rigid fixation. Combining sternal approximation with fixation may improve sternal healing.

Objective. To report a randomised first-in-man study evaluating sternal healing, postoperative pain and complications following sternal closure using either WC or a novel device (SL-360) combining a cerclage band with a rigid plate.

Methods. Patients undergoing elective cardiac surgery were randomised to SL-360 ($n=26$) or WC ($n=24$). Independent radiologists evaluated 3- and 6-months postoperative computed tomography (CT) scans for sternal healing. Healing scores and rate of sternal union were evaluated along with pain, complications and proportion of pain-free patients up to 6-months. Logistic regression explored the relationship between healing and pain.

Results. At 3-months, sternal healing was greater in SL-360 compared with WC (69% v. 25%; $p=0.002$). The mean (standard deviation (SD)) CT scores at 3 months were 3.3 (0.9) for SL-360 and 2.2 (1.0) for WC ($p=0.001$), with an improved trend at 6 months ($p=0.055$). The difference in pain scores following exertion/coughing reached statistical significance at one-month postoperatively, favouring SL-360 (0.9 (1.0) v. 2.2 (2.0); $p=0.007$). Logistic regression demonstrated that improved healing leads to a higher probability of being pain-free after exertion (OR 2.3; 95% CI 1.2 - 2.2; $p=0.009$). Superficial wound infections occurred in 8.3% ($n=2/24$) and 3.8% ($n=1/26$) of WC and SL-360 patients, respectively ($p=0.6$).

Conclusion. Sternal closure combining rigid fixation combined with cerclage results in improved sternal healing, and a significant improvement in postoperative pain.

A new technique of scapulothoracic fusion using a 3D-printed customised scapular titanium implant

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Introduction. Scapula alata (SA) is a rare, debilitating condition caused most commonly by the palsy of the trapezius or serratus anterior muscle as result of traction or compression of the spinal accessory or long thoracic nerve, local inflammatory or tumoural disease, or traumatic or iatrogenic nerve injury. Chronic shoulder pain, muscle weakness and inability to elevate the affected extremity above shoulder level are frequently associated symptoms, thus quality of life is significantly affected. When conservative management with physical therapy fails in achieving pain relief and restoration of joint range of motion, surgery may be indicated. Current surgical options include a variety of dynamic muscle transfers which offer the theoretical advantage of improving range of motion, but are associated with high morbidity, high clinical recurrence rate and potential adverse effects on the donor site or static stabilisation procedures.

Methods. We present a video and describe in detail a new technique of scapulothoracic fusion using a 3D printed customised scapular titanium implant that was used in a patient with painful lateral winging of the scapula. A customised multi-perforated 3D-printed titanium implant and multi-filament steel cerclage cables were used to assure a strong stable scapulothoracic bone union and were found of paramount importance for success.

Results. Postoperatively, patients were immobilised with a sling for 5 weeks although dangling activity to avoid stiffness in the elbow and wrist, as well as hand motion, was recommended. At 12 weeks, the patient was able to resume normal activities and reported a significant improvement in the shoulder function, release of the pain and high satisfaction rates on follow-up.

Conclusion. The use of 3D-printed customised scapular implants has made scapulothoracic arthrodesis safer, faster and easier than previous fusion techniques and offers undoubted benefits for patients with SA. When comparing with alternative dynamic muscle transfer techniques, this fusion method allows for more predictable results, is easily reproducible and provides a rapid functional recovery allowing a rapid return to daily activities.

Minimally invasive repair of pectus carinatum following the Pérez technique in a patient with Marfan syndrome and severe deformity

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Introduction. Marfan syndrome is a primary hereditary connective tissue disorder as result of the dominantly inherited deficiency of fibrillin-1. Up to 70% of patients with Marfan syndrome present with

congenital chest wall deformities including pectus excavatum (PE) and pectus carinatum (PC) although mixed forms of deformities or asymmetry due to abnormal unbalanced growth of the costal cartilage or rotation of the sternum are also frequently present. In cases of PE, symptoms do not differ from those present in non-syndromic patients, while in cases of PC indication of corrective surgery is mostly cosmetic. **Case.** We report the case of an extremely severe PC deformity in a patient with Marfan syndrome leading to low self-esteem and social anxiety problems. For these reasons, a corrective surgery with a recently developed reverse Nuss method (MIRPC by the Pérez technique) was indicated. Steps of the surgical procedure are shown in detail and images of the result are presented. As for the standard Nuss procedure for PE, bilateral 4 cm-long incisions were performed at the anterior axillary lines. A pre-shaped curved steel bar was inserted intrapleurally and advanced until reaching the opposite pleural space, with only its medial segment running above the sternum, thus pushing the protrusion back. The bar was then fixed to the underlying ribs with wires and incisions were closed without drains

Conclusion. By the use of this new corrective procedure, excellent remodelling of the chest wall was achieved in absence of anterior visible incisions. In contrast to the Nuss procedure for PE, the advancement of the introducer above the sternum instead of between the sternum and the heart, makes the surgery easier and safer for surgeons.

The sternal transection as methods to improve outcomes of the Nuss procedure in adult patients with high chest wall rigidity

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Introduction. The Nuss procedure for pectus excavatum provides excellent thorax remodelling in children and adolescent patients with malleable deformities. However, in cases of older adolescent or adult patients with higher chest wall rigidity, the bar can lead to severe or prolonged postoperative pain or can fail to completely elevate the depressed sternum. Additionally, the post-surgical increased mechanical tension of the chest wall as result of the interaction between rib cage and implant is thought to be responsible for the higher bar dislocation rates found in this subgroup of patients.

Methods. On the basis that reducing chest wall stiffness before introducing the implant bar can improve outcomes of the Nuss procedure in patients with rigid rib cage, an endoscopic guided sternal transection 5 cm below the level of the Louis angle was performed on 20 consecutive adult patients with PE. Surgical steps are shown in detail and results of the surgery are presented and compared with those of patients operated in a standard manner.

Results. Better correction of the deformity in the immediate postoperative period was achieved in the group who underwent sternal transection, while postoperative pain intensity decreased in this group. Satisfaction with the cosmetic results of the operation was very high and similar in both groups. Two dislocations occurred only in the group of adult patients who underwent the standard procedure.

Conclusion. Performing a transverse sternotomy is easy and safe and can be achieved through only an additional small incision. It significantly reduces the rigidity of the rib cage, thus facilitating the immediate correction of the deformity and decreasing the postoperative pain.

Use of transoesophageal echocardiography for enhanced safety during bar removal procedures after minimally-invasive repair of pectus excavatum

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Introduction. Hardware removal, after minimally-invasive pectus excavatum repair, has been rarely associated with life-threatening haemorrhage from the heart, aorta, internal mammary arteries, and/or lung. Currently, there is no generally-accepted standard intraoperative monitoring technique for these procedures. We hypothesised that the use of transoesophageal echocardiography (TEE), currently available in most modern operating room suites offering cardiac surgery and commonly used by many anaesthesiologists, during Nuss bar removal, could enhance safety of the procedure without producing complications.

Methods. IRB-approved, retrospective review of all patients who underwent Nuss bar removal with intraoperative TEE monitoring over a 4-year period, from March 2013 to May 2017, was completed. Bar removal procedures were done in the supine position, under general anaesthesia. Bars were removed after being straightened and gently shifted slightly from side to side, while TEE images were monitored by both the surgeon and anaesthesiologist. Distortion of the cardiac silhouette, new pericardial effusion, and/or cardiac arrhythmias would be considered evidence of possible bar adherence and/or injury to the heart and would have triggered immediate conversion to sternotomy or thoracotomy.

Results. A total of 87 consecutive patients, with a mean age of 20 years (range 11 - 33), 73.5% male, were identified. Two bars were placed in 75 of the 87 patients. Bars had been in place for a mean duration of 30 months (range 4 - 83). Bar removal procedures took an average of 67 minutes (range 28 - 309). TEE gave excellent visualisation of the cardiac silhouette and pericardium in all cases, with no further complications or oesophageal injuries. No patient required insertion of an intra-arterial monitoring line, a postoperative chest X-ray, or overnight hospitalisation after bar removal. Patients were discharged from the recovery room at an average of 89 minutes after completion of surgery.

Conclusion. TEE appears to offer a minimally invasive, safe way for the surgeon and anaesthesiologist to visualise the pericardium and its contents during Nuss bar removal. Significant cardiac or mediastinal injuries should be immediately visible. Since many hospitals already have TEE equipment, its use appears to be cost-effective, allowing surgeons to safely discharge patients almost immediately, without overnight hospitalisation.

Staged chest wall reconstruction in thoraco-omphalopagus conjoined twins

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Objective. To describe successful chest wall reconstruction in a challenging conjoined twin case, incorporating components of multiple well-established surgical techniques previously described for the treatment of pectus excavatum/carinatum and bifid sternum.

Methods. Twins A and B, diagnosed *in utero* by prenatal ultrasound and MRI studies, were delivered at 34 weeks' gestation (combined weight 4.3 kg). Bilateral subcutaneous lateral abdominal wall tissue expanders were placed and gradually enlarged as outpatient procedures. At age 6 months, separation was successfully completed, with division of their shared hepatic parenchymas and gastrointestinal tracts. Both hearts shared a common pericardial sac. Following separation, multiple intraoperative attempts at sternal closure resulted in acute hypotension secondary to cardiac compression. Therefore, acellular dermal matrix (Alloderm) patches were sewn to the sternal edges and covered with large skin flaps, resulting in a type of bifid sternum appearance and significant anterior protrusion of the sternal halves similar to pectus carinatum. Over the subsequent 10 years, the children each had rigid plastic chest wall protector plates created and then revised, so that they could participate actively in competitive soccer and other sports.

At age 11 years, sternal closure (and carinatum correction) was achieved in each twin using a Ravitch-type approach: midline anterior chest wall incisions were made, pectoralis muscles were reflected laterally, sub-perichondrial resections of cartilages 2 - 7 were performed, inverted V-shaped manubrial wedge osteotomies were performed, posterior sternal periosteum was approximated, and then sternal halves were brought together with figure-of-8 steel cables.

Results. Reconstruction operations for A and B took 6 and 4 hours, respectively. Twin A developed mild superficial wound breakdown, which was successfully treated with vacuum-assist (Wound Vac) and, at 2 months, removal of one exposed sternal cable. Two years following reconstruction, A and B have closed wounds, excellent chest wall stability, and no limitations of activity. Breast reconstruction is being planned.

Conclusion. Staged chest wall reconstruction after successful separation of thoraco-omphalopagus twins, can be achieved using modifications of multiple established techniques, including use of dermal matrix, sub-perichondrial cartilage resections, wedge sternal osteotomies, and sternal cables.

Commercially-produced, surgeon-directed customised titanium implants for chest wall reconstruction for complex Poland's syndrome

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Introduction. This study describes a commercially-produced and customised titanium chest wall implant for reconstruction of complex Poland's syndrome. Computer-aided design software and web-based conferencing allowed the implant to be designed and modified by the patient's surgeon and then manufactured by a commercial vendor.

Methods. A 12-year-old female was born with absence of the right breast, pectoralis muscles, latissimus dorsi, anterior aspects of ribs 2 - 10, and a shortened hypoplastic sternum. Fluoroscopy showed no diaphragmatic movement abnormalities. Computerised tomography (CT) showed a hypoplastic hemithorax with significant pulmonary hypoplasia. Pulmonary function tests (PFTs) were 47 - 50% of predicted. Since radiographic bone age studies showed advanced bone age, with minimal anticipated remaining growth, reconstruction was planned.

Chest CT (1 mm thickness) with shaded surface display (SSD) images were obtained. Design was coordinated during 2 brief web-based video conferences using commercially-available software (Cisco WebEx, Milpitas, CA) with the surgeon, a design engineer (3D Systems, Littleton, CO), and a development engineer (Zimmer/Biomet, USA). After design approval, a prototype plastic model was used to create a customised five-piece titanium implant for reconstruction of the hypoplastic sternum and five adjacent ribs. The implant was created under the United States Food and Drug Administration (FDA) Custom Device Exemption (CDE) law, modified in 2012 by the FDA Safety and Innovation Act (FDASIA).

Results. After dual-lumen endotracheal tube placement and right arm elevation using finger traps, a sub-mammary incision was made. Underdeveloped chest wall cartilages were excised. An extrapleural dissection improved lung mobilisation/expansion. The first rib was incised to allow slight sternal derotation. The sterilised plastic prototype model was used to check implant fit. The implant was secured with 60 titanium screws. She was extubated at the conclusion of the operation, discharged home on the fifth postoperative day, and returned to school and full activity within four weeks. Breast implant placement is planned for the future.

Conclusion. Advances in 3-dimensional design software, web-based conferencing, and titanium manufacturing now allow chest wall surgeons to design, modify, and implant customised multi-piece implants for chest wall reconstruction in patients with complex Poland's syndrome.

The psychological impact of chest wall defects

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Introduction. Pectus excavatum (PE) has popularly been believed to have only cosmetic impact. Recognition that PE can have a negative impact on body image, has received scant investigation of the psychological impact on patients.

Objective. To report outcomes of an on-going prospective study exploring patient experience(s) before and after surgical treatment.

Methods. A sample of 10 patients referred for corrective surgery were invited to take part in a two-part psychology interview that utilised Interpretative Phenomenological Analysis to explore opinion and experiences of living with PE pre- and post-surgery.

Results. Pre-surgery themes include the difficulty being taken seriously by health professionals and family, in handling other people's (imagined) reactions, and in getting support. PE impacted on their careers: patient 1 and 2 avoided careers because of their PE and patient 3 was on long-term sick leave. All participants had searched for information online and many had watched videos of operations as part of their preparation. Hopes for what the operation would do for them were all about regaining a quality of life that they had lost. None had unrealistic expectations. They just wanted to go from 'hole' to 'whole'.

Post-surgery themes include problems with hospital administration, short notice for operations and also sudden cancellations. Learning about their new body was encapsulated by the phrases 'I have an armpit' and 'my body is like a weird different body'. All reported positive impacts, including one person who had experienced postoperative infection that required 6 months of treatment, noting huge improvements in quality of life. All would recommend the surgery. All wanted provision of more information prior to surgery. All were enjoying their new bodies and summed up the experience as 'I mind the scar less than what was before'.

Conclusion. These themes will be refined as analysis progresses and further interviews are conducted. Key take home messages are that PE is not just cosmetic and that we as health professionals need to do a better job at recognising this.

Chest wall Ewing sarcoma, improving outcomes for a rare cancer in Egyptian adolescents and young adults

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Objective. To evaluate the multidisciplinary management modalities for chest wall Ewing sarcoma/primitive neuroectodermal tumours (ES/PNET) and its outcomes as regard short-term relapse and survival to compare our results with the observations of other intergroup studies across the world and to recommend the best approach for the treatment.

Methods. Our retrospective study was conducted on all patients who presented with thoracic ES/PNET at the National Cancer Institute, Cairo University, from January 2000 to December 2015.

Results. A total of 30 patients were assigned to two groups. The 1st group consisted of patients who received standard neoadjuvant chemotherapy, followed by local treatment and adjuvant chemotherapy every 3 weeks. The second group had received interval-compressed neoadjuvant chemotherapy followed by local treatment and adjuvant chemotherapy every 2 weeks. Local treatment in the form of surgery, i.e. complete, full-thickness chest wall excision and immediate reconstruction was performed for all cases in the study, followed by radiotherapy in 12 selected cases. In general, over a median follow-up period of 114 months, the 3-year overall survival for the patients with metastases at presentation was 10% v. 41% of patients without

metastases at diagnosis ($p=0.007$). In non-metastatic patients, interval-compressed chemotherapy has improved the 3-year overall survival rates from 48.5% in the first group to 83.3% in the second group ($p=0.02$). It also had improved the 3-years event-free survival rates from 30.8% in the first group to 53.6% in the second group ($p=0.262$).

Conclusion. With extended follow-up, our series demonstrates that upfront interval-compressed multi-agent chemotherapy followed by local control in the form of adequate surgery, alone or combined with radiotherapy, along with concomitant chemotherapy followed by adjuvant chemotherapy are the backbone in the treatment of patients with chest wall ES. Our study has limitations, but with a rare disease like this, randomised data are scarce. However, our experience spans two decades during which chemotherapy, surgical technique, and radiotherapy methods changed dramatically. Despite these shortcomings, our study represents one of the largest single-institution series addressing chest wall ES.

Investigation of causes of exercise dysfunction of pectus excavatum with cardiopulmonary exercise testing: A two-centre study

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Introduction. Causal mechanisms of reported symptoms of exercise in patients with pectus excavatum (PE) remain open to investigation. Cardiopulmonary Exercise Testing (CPET) has shown early promise as a test that might elucidate the cause of exercise dysfunction. **Objective.** We have undertaken a two centre collaborative study to evaluate whether previously reported findings are confirmed by more comprehensive studies.

Methods. Between 2006 and 2018, adult patients requiring surgical correction of PE underwent pre-operative assessment of anatomical and physiological status, by CT Scan, spirometry and CPET. CPET measurements recorded at anaerobic threshold and peak exercise were work, oxygen utilisation and characteristics of ventilation. Data are presented as percentages of predicted values, means and standard deviation (SD). Statistical analysis used students t-test, $p < 0.05$, univariate and multi-variate regression analysis to identify predictors of exercise function.

Results. A total of 345 patients (76.0% male), with a mean (SD) age of 29.2 (11.0) years, had Haller's indices on CT scan of 4.45 (1.73). Results of CPET demonstrated that, at peak exercise, the mean (SD) heart rate (HR) was 85.3% (8.7), and work achieved was 85.6% (22.0), while 94.4% of patients achieved respiratory equivalents (RER) greater than 1.1 (mean (SD) 1.22 (0.11)).

Oxygen delivery as measured by VO_2 max and VO_2 anaerobic threshold (AT) was reduced: 73.2% (15.6) (normal $>80\%$) and 41.1% (12.3) (normal 40% - 60%), respectively. O_2 pulse, a surrogate of stroke volume, was 86.0% (16.6) (normal $>80\%$). Measures of ventilation equivalents (VE BTPS) were reduced to 41.2% (11.7) (normal $>85\%$) and breathing reserve (BR%) was elevated at 46.9% (14.8) (normal

15%). O_2 pulse, showed correlation with BR% (-0.39), VO_2 AT (0.57) and VO_2 max (0.87) ($p=0.0001$). Multiple linear regression identified ventilation equivalents (VE BTPS), respiratory rate and O_2 pulse as composite predictors of VO_2 max ($R^2 = 91\%$; $p=0.001$). VE BTPS and end-tidal CO_2 were predictors of O_2 pulse ($p=0.001$).

Conclusion. This study confirms a relation of PE-related symptoms to compromised chest wall mechanics and cardiorespiratory dysfunction. The results suggest causation is attributable to compromised mechanisms of ventilation and support the premise that corrective surgery should achieve restoration of chest wall mechanics.

A modified classification of flail chest

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Introduction. The current definition for flail chest fails to recognise the complexity of chest injuries. Comprehensive planning of the management of chest trauma is therefore impeded with a potential of increased injury related mortality.

Objective. We report a classification of chest wall injuries that encompasses the complexity of thoracic wall injuries and facilitates planning of therapeutic strategies.

Methods. Our institute introduced chest wall reconstruction in August 2014 as part of a strategy for management of polytrauma patients. One hundred patients underwent chest wall reconstruction following major trauma. Evaluation of the patterns of injury allowed formulation of this classification. A retrospective review has been performed to define the distribution of injuries witnessed in patients.

Results. The average age of patients was 56.8, 70% were male and road traffic collisions were responsible for 54% of injuries. The incidence of these injuries was as follows: type A ($n=20$), B ($n=26$); C ($n=18$); D ($n=3$); combination ($n=11$); and no-flail ($n=22$). Five patients (25%) with a Type A flail also sustained a clavicular fracture, while 17 (85%) suffered a sternal fracture contributing to the flail segment. In this group (A) 4 (20%) patients required sternal fixation to stabilise the flail segment. Within the type D group, 2 (66%) patients sustained both thoracic vertebral fractures and scapula fractures. The New Injury Score was 36.7 (34.5 - 39.01) indicating severe polytrauma.

Conclusion. We believe the new classification for flail chest facilitates appropriate assessment and management of patients with major chest wall injury. These patients have life threatening injuries as demonstrated by high NIS scores, thus enhanced assessment and definitive management of such injuries will optimise patient outcomes.

Understanding cardiopulmonary exercise testing in relation to pectus carinatum: Exercise physiology

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Introduction. Patients with pectus carinatum are a minority among patients with congenital chest wall abnormalities. The defects can be

quite severe with prominent protrusion of the sternum and chest wall. It is most commonly considered to carry no physiological impact and only primarily cosmetic importance.

Objective. To present the characteristics of exercise physiology as documented by cardiopulmonary exercise testing (CPET) in order to elucidate whether the defect truly has an impact on exercise.

Methods. We will provide a discussion of cases of patients who have pectus carinatum who have been investigated with the standardised protocol of our Institute, i.e 2D computed tomography scan and 3D reconstruction, as well as comprehensive cardiopulmonary exercise test evaluation. Selective use of dynamic pulmonary function tests is also referenced. I will aim to answer the following questions: Do patients with pectus carinatum experience exercise dysfunction? and How do I identify the cause of this exercise dysfunction and the relationship to the defect?

Results. I will undertake this presentation by deconstruction of detailed CPET reports and present a structured and stepwise pattern of interpretation of the data. I will examine by evaluation of heart rate, RER and metabolic status to identify if patients apply themselves maximally to exercise. With markers of work, VO_2 max and VO_2 anaerobic threshold I will present evidence of efficiency of exercise performed. Causal relationship to cardiac and respiratory function will be evaluated by deconstruction complex respiratory and cardiovascular changes, as in a similar presentation regarding Pectus Excavatum. It is the aim of this presentation to deliver a foundation of knowledge that will enable comprehension of the impact of pectus carinatum and facilitate interpretation of cardiac and respiratory

Conclusion. I will provide information regarding the impact of pectus carinatum on exercise capacity, and present theories for the changes noted. It is intended that this presentation will challenge current perception that holds that the defect does not cause exercise dysfunction.

Back to basics: Cardiopulmonary exercise testing – normal expectations of exercise

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Introduction. Cardiopulmonary exercise testing (CPET) provides assessment of exercise responses involved in the pulmonary, cardiovascular, haematological, neuropsychological, and skeletal muscle systems, which are not adequately reflected in the measurement of individual organ system function alone. This non-invasive dynamic physiological overview allows the evaluation of both submaximal and peak exercise responses providing relevant information for clinical decision-making. CPET is increasingly being used in a wide range of clinical applications for the evaluation of diagnosed and undiagnosed exercise intolerance. It provides objective measures of functional capacity and impairment.

Objectives. To provide a learning foundation for a second presentation regarding changes that are found in relation to chest wall defects.

Methods. Peak exercise capacity is defined as ‘the maximum ability of the cardiovascular system to deliver oxygen to exercising skeletal muscle and of the exercise muscle to extract oxygen from the blood’. Subsequently, exercise tolerance is determined by three factors: pulmonary gas exchange, cardiovascular performance (peripheral vascular tree) and skeletal muscle metabolism. Oxygen delivery may be represented by the formula:

$$VO_2 \text{ max} = (SV_{\text{max}} \times HR_{\text{max}}) \times (CaO_2 \text{ max} - CvO_2 \text{ max})$$

Results. It is the intention of this presentation to describe the methods used to undertake CPET and to present in a step-wise manner the interpretation of normal exercise function on CPET, and interpretation of the elements of exercise that influence oxygen delivery. We will discuss what the changes are in cardiac, respiratory and metabolic status that might be expected in a patient with normal exercise function. We will examine what might be considered normal patterns of data and graphical changes of CPET. Examples will be presented of patients cardiac or respiratory pathologies as a means of exemplifying normal patterns.

Conclusion. It is intended that through this presentation a clinician will gain a foundation of knowledge in regard to CPET that allows effective use of this investigation for clinical and research purposes.

Chest wall trauma

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Introduction. The chest wall has both a structural and functional purpose. Structurally, it functions as a barrier against external forces that may harm the respiratory, circulatory, gustatory and lymphatic organs. Its compliance allows the absorption of forces that will affect the function of the underlying protected organs. Functionally, it plays a role in the mechanism of breathing by means of expansion and retraction as well as generating a negative intrapleural pressure that prevents collapsing of the lungs.

Chest wall trauma is one of the most commonly encountered injuries associated with high energy trauma. Statistical evidence on the incidence of chest wall trauma is sparse due to insufficient data-capturing protocols. As a result, accurate figures for South African case studies are scarce. However, it is well known that incidences of chest wall trauma are significant in South Africa. Generally higher income countries gather more statistical data regarding injury demographics. These statistics are more readily available online and was referred to in this discussion.

Chest wall trauma can be subdivided into blunt and penetrating thoracic trauma, respectively. Understanding the physics and pathophysiology pertaining to these types of chest wall trauma allows for prompt identification and treatment of other associated injuries.

The discussion gives an overview of the anatomy and physiology of the chest wall, available epidemiology concerning chest wall trauma, the physics and pathophysiology of chest wall trauma as well as associated injuries. The diagnosis and management of chest wall trauma have also been addressed.

A rare, giant extraocular, anterior chest wall sebaceous carcinoma

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Introduction. Sebaceous carcinoma is a rare aggressive cutaneous malignant tumour. It accounts for less than 1% of all cutaneous malignant tumours. Sebaceous carcinomas are divided into ocular and extraocular constituting 75% and 25%, respectively. The most common extraocular site is parotid gland. Chest wall is a rare site of this tumour.

Case. We report a case of 45-year-old African male who presented to our unit with 2 years history of a large right anterior chest wall tumour. He reported that the tumour started as a small lump, which grew gradually over two-year period. The patient reported no history of visceral malignancy and radiation exposure. However, his risk factors were age and immunosuppression in the form of Human Immunodeficiency Virus (HIV). The tumour was initially diagnosed as sebaceous adenocarcinoma by incisional biopsy before he was referred to us a year ago. Excision of the tumour with wide margins was undertaken and histology confirmed sebaceous carcinoma.

Conclusion. Sebaceous carcinoma is a rare aggressive malignant tumour originating from sebaceous gland. Early diagnosis and wide excision with negative margins improve survival.

Use of artificial intelligence to improve diagnosis and care planning of patients with pectus excavatum

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Introduction. Pectus excavatum (PE) is the most common chest wall defect around the world with a frequency of 1:400. PE patients may experience shortness of breath, lack of endurance, chest pain, or fatigue. These symptoms are inconsistent in childhood and may worsen with increasing age. If uncorrected, some patients will develop cardiopulmonary symptoms as adults and others will experience a worsening of already existing, but mild symptoms. Since a misconception of PE as a 'cosmetic disorder without physiologic consequences' persists, the use of artificial intelligence (computer-aided diagnosis, especially Blockchain) aims to improve diagnosis and care planning of PE patients.

Objective. We propose to develop an application (app) for smartphones using different collective intelligence protocol, aiming to improve diagnosis of PE, care planning and information of PE patients.

Results. The app handles an extensive amount of information such as biometric data, symptoms, and form, depth and photos of PE. The app provides information not only to physicians, but also to patients and other professional groups such as physiotherapist, and ensure

experience exchange between the involved persons. In case of questions or problems, the patient may contact its physician directly. The app helps to monitor and to adapt the therapy (i.e. application time for the suction cup; quality of life; consumption of painkillers).

Conclusion. Treatment recommendations are based on the use of artificial intelligence and of Blockchain technology that allows secure data sharing between patients and physicians with accurate traceability. This technology aims to enforce collective intelligence into a network of stakeholders and to warrant the most appropriate medical decision.

Our bar placement strategy for the Nuss procedure: Pinpoint targeting using intraoperative rib-marking with the aid of 3D computed tomography imaging

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Introduction. Accurate Nuss bar placement is key for successful pectus excavatum repair. We use 3D computed tomography (CT) imaging along with rib marking during operation and prick-guidance on the hinge point for this purpose. To obtain the best position, our strategy and the procedures are introduced.

Objective. To facilitate preoperative planning of the procedure.

Methods. For preoperative chest evaluation, we routinely take CT with 3D image. We simulate the operative procedure on the computer screen, and bar position and hinge points are determined. With this information, rib contours and hinge points were marked on the skin using a marker pen before the operation. Under the thoroscopic guidance, we make a prick with a fine needle from the hinge point mark on the skin and an introducer is penetrated from this point. With crane technique and careful mediastinal dissection, contralateral thoracic cavity is approached. Under the same thoroscopic vision from the right side through the mediastinal window, a fine needle is pricked from the left hinge point. The introducer penetrates the point, then guide tape is passed through.

Results. With these procedures described above, it becomes clear how the sternum and the ribs are elevated by the bar(s). To decide the bar position and the hinge points preoperatively with the aid of 3D CT, we can imagine the whole operative process and the postoperative shape.

Conclusion. 3D CT imaging assists in the operative procedure for repair of pectus excavatum.

3D body surface scanning index for the evaluation of chest deformity in patients with pectus excavatum

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Introduction. Evaluation of chest deformity in patients with pectus excavatum (PE) using 3D body scanning has become a new modality.

With this new technique, we analysed a new index, the external correction index (E-CI).

Objective. We evaluated and compared E-CI with Haller index (HI) and correction index (CI).

Methods. Seventy-eight PE patients (60 males, 18 females) underwent 3D body scanning using the Structure Sensor. For scanning, patients were laid on an operation table under general anaesthesia before the Nuss procedure. The data were analysed using 3D-Rugle imaging software. E-CI was measured using 3D tomographic imaging at the deepest point. The length at the deepest point from the back (A) and the length at the high point (B) were measured. E-CI was calculated as follows: $(B-A)/B \times 100$ (%). HI and CI were measured on preoperative CT image.

Results. The median age of 78 cases was 15.6 years (range 4 - 45). The median HI was 6.4 (range 3.0 - 43.1), median CI 41.4% (range 17.3 - 89.6) and median E-CI was 14.0% (range 4.2 - 28.8). The correlation between E-CI and HI was 0.54 ($p=0.0001$) and between E-CI and CI it was 0.76 ($p=0.0001$).

Conclusion. The body surface 3D scanning is an accurate, radiation-free method for evaluating PE deformity. E-CI is well correlated with conventional indices. The 3D technique holds the potential as a standard methodology to evaluate PE in the future.

Repairing chest-wall deformation in children

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Introduction. Pectus carinatum (PC) is a complex congenital anatomic and cosmetic defect of development in children aged 4 - 18 years. PC can slow physical growth and have psychological effects.

Objective. To determine the efficiency of conservative and surgical repair of PC in children.

Methods. Since 2015, 611 (425 (69.5%) boys and 186 (30.5%) girls) children with PC have received care and follow-up in our Paediatric Orthopaedic Department. Following Russian authors' and H J Park of South Korea's categorisation system, the following PC types were classified: monubriocostal ($n=125$; 20.5%); corpocostal ($n=199$; 32.5%); and rib type ($n=287$; 47.0%). By level, the deformation was divided into: I-level ($n=102$; 16.7%); II-level ($n=281$; 46.0%); III-level ($n=228$; 37.3%). The symmetric type was seen in 331 (54.2%) children, asymmetric in 216 (35.4%); mixed form (Park's classification) in 46 (7.5%) and upper type in 18 (2.9%). All patients passed complex examination, and 534 (87.3%) were treated conservatively and surgically. Evidence for conservative treatment was cosmetic defect and intrathoracic pressure ≤ 10 kg, following Yuksel in Turkey, and intrathoracic pressure ≤ 17 kg and cardiac insufficiency in patients in III-level. A total of 461 (86.3%) patients were selected for conservative treatment, using Yuksel's orthosis, FMF by M F Marcelo (1992) or orthosis made in Uzbekistan. Evidence for surgery was high intrathoracic pressure (≤ 10 - ≥ 25 kg, following Yuksel), high thorax rigidity and a deformation not correctable by conservative means, mostly in children aged 15 - 18 years and older. Operative treatment was used in 73 (13.7%)

patients: 44 (63%) with Abramson's method; 27 (37%) the Sandwich-II technique; and 2 (2.7%) Ravich's method.

Results. On follow-up after 6 - 12 months, it was found that: after conservative treatment 408 (88.5%) children had 'good' results; 48 (10.4%) had 'satisfactory' results; and in 5 (1.1%) it was suggested they continue to wear the orthosis. Following operative repair, 50 (68.5%) had 'good' results; 18 (24.7%) were 'satisfactory'; and fixing of wire movement was needed in 5 (6.8%). In these cases, a second surgery was done successfully. Levelling of the thorax and improvements in the cardiorespiratory system were observed. Moreover, satisfactory cosmetic repair of the defect, and psychological improvements, were noted.

Conclusion. The treatment of PC in children using the suggested methods gave us a high percentage of positive results.

Treatment of thoracic kyphosis in children

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Introduction. Kyphosis is a complex congenital malformation of spinal curvature resulting in sagittal flatness of back convexity.

Objective. To determine the efficiency of conservative treatment of kyphosis in children.

Methods. A total of 237 children with thoracic kyphosis were under observation and received treatment: 149 (69%) boys and 88 (30.5%) girls, aged 4 - 18 years. Congenital kyphosis was found in 12; derived in 19; postural in 52; rachitic in 8; and mobile or compensatory in 146 patients. According to degree of deformation, they were classified as follows: I degree, 57 (16.7%); II degree, 162 (46.0%); III degree, 18 (37.3%). Thoracic localisation was observed in 209 (54.2%), cervical in 22 (35.4%) and lumbar in 6 (7.5%). In combination with other diseases, it was found with pectus carinatum (PC) in 183, pectus excavatum (PE) in 43 and kyphoscoliosis in 11 patients. Evidence for conservative treatment of patients was intrathoracic compression, intervertebral nerve injuries and cosmetic defect with intercostal damage. With conservative methods, gentle correction of posture was used in 194 (86.3%) patients, and gentle correction and compressive orthosis in cases of combination PC and compressive orthosis. Evidence for operative treatment of kyphosis was III degree kyphosis, an angle $>70^\circ$ from total type and deformation that is uncorrectable by conservative means, especially in children aged 15 - 18 years and older. A total of 13 (13.7%), and 7 patients with kyphoscoliosis, were sent to vertebrology for further treatment.

Results. At follow-up 3, 6 and 12 months after conservative treatment, 147 (88.5%) children had good results, 32 (10.4%) were satisfied, and 15 (1.1%) patients were advised to continue wearing the spine orthosis. Levelling of the spinal column and chest form were observed, and improvement in the function of intrathoracic organs and of intervertebral nerves. Satisfaction with the cosmetic treatment of the thoracic defect and psychological improvements in patients were noted.

Conclusion. The treatment of kyphosis in children according to the suggested methods provided a high percentage of positive results.

Repair of concave-type rib deformation of the chest in children

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Introduction. Concave rib deformation (protrusion) of the chest is a frequently observed rib defect in children.

Objective. To examine the development of complex conservative actions with the administration of Eckart Klobe's vacuum bell (2006, Germany) and operatively setting plates by cross-type (Park, South Korea) on concave ribs deformation.

Methods. In the Paediatric Orthopaedic Department of State Children Multibranch Medical Center, Samarkand, Uzbekistan, 1 504 children with rib deformation and rib-defect development have been examined since 2015, aged from 4 to 18 years. A total of 60 (4%) children aged 4 - 18 were found with concave rib deformation of chest: of these, 41 (68.3%) were observed to have the one-sided type, and 19 (31.7%) two-sided. All patients received complex examination in hospital. Conservative and operative treatment were carried out in 47 (78.3%) patients, and 13 (21.7%) patients had concomitant diseases. Of these, 11 (23.4%) patients were recommended for operation cross-type using Park's methods (at ages 16 - 18 years), and 36 (76.6%) to use Haecker's vacuum bell method. Evidence for conservative treatment was cosmetic defect, not very high resistance or flexible rib and impression in lower part of chest.

Results. Evidence for surgery of rib protrusion was high rigidity and resistance of the deformed part, and being unable to correct by conservative means using the vacuum bell, especially in children 16 - 18 years and older. Step-by-step conservative treatment with the help of the vacuum bell in 36 (76.6%) showed that after 3, 6 and 12 months, respectively, 28 (77.9%) patients had good results, 4 (11.1%) were satisfied, 2 (5.5%) were recommended to continue wearing the vacuum bell and 2 (5.5%) patients with unsatisfactory results were recommended the cross-type operation at 16 - 18 years. After the repair, the levelling of ribs and chest form, satisfactory cosmetic repair of the defect and psychological improvement in patients could be observed.

Conclusion. The treatment of rib protrusion in children by the suggested methods yielded positive results.

Back to basics – the radiology of pectus excavatum

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Introduction. Pectus excavatum is commonly considered to be a defect with a limited spectrum of anatomical abnormality. Yet clinical experience of its management clearly causes realisation of the wide variation in structural patterns of the defect. These differing patterns impact on both symptoms and surgical strategies of management.

Objective. To provide a discussion of anatomical characteristics evident radiologically.

Methods. I will provide discussion of the range of radiological investigations, chest X-ray, 2D and 3D computed tomography scan imaging, that are available to determine the structural anatomy of pectus excavatum. I will provide a discussion regarding the impact of the defect on the thoracic cavity as a whole, discussing the bony and cartilaginous defects that are evident. Discussion will focus on the classification previously reported by our institute. I will in addition provide discussion as to other anomalies of the chest wall that may present as pectus excavatum. The aim of my talk is to provide a foundation of understanding that will facilitate comprehension of the impact of this problem on cardiorespiratory physiology and clinical management.

Results. Discussion of detailed skeletal changes of types A, B and C pectus excavatum will be provided. The poor relationship to the Haller Index will be explored and the additional importance of the impact on total chest volume will be addressed. The relationship of structural changes, and the characteristic of bony deformity will be addressed, allowing considerations that are required during surgical correction.

Conclusion. I wish to provide a working understanding of the radiological characteristics of pectus excavatum and related defects that will allow clinicians to plan assessments and treatments.

Sternal suspension technique in treatment of pectus excavatum after open repair of congenital heart disease

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Objective. The purpose of this study was to evaluate the efficacy of using sternal suspension technique in correcting pectus excavatum after open repair of congenital heart disease and improve the safety of this technique.

Methods. A retrospective study was conducted for 8 cases of pectus excavatum (5 males and 3 females) treated with sternal suspension procedure after open repair of congenital heart disease from October 2005 to October 2018 in our institution. The median age of the patients was 5.75 years (range 3.75 - 12.33) and the median Haller index was 4.20 (range 3.60 - 5.19). All patients underwent cardiac ultrasound and chest computed tomography scan to evaluate the severity of retrosternal adhesions.

Results. All patients underwent the sternal suspension procedure successfully with a median operation time of 55 min (range 30 - 230 min) and blood loss was minimal. One patient suffered pneumothorax after operation. The complication rate was 12.5%. The patients were followed up for 6 months to 13 years, with an excellent and rate of 100%.

Conclusion. Sternal suspension technique is a safe and effective procedure for pectus excavatum after open repair of congenital heart disease. To improve the security and reduce the incidence of cardiac injury, the sternal suspension technique is a promising alternative for pectus patients with severe adhesions after open repair of congenital heart disease.

The psychological and psychiatric impact of pectus malformations in a South African context

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Individuals with pectus malformations, including excavatum and carinatum, may experience psychological distress, including altered body image, low self-esteem, embarrassment and stigma. Although physiological parameters are usually considered as the determinant for intervention, these psychological aspects impair the quality of life of affected individuals.

There may be an impact on the individual's body shape, leading to altered posture and wearing loose clothing. There is avoidance of certain activities, such as swimming or travelling. Concerns with regards to heritability of the malformation has been documented. Bullying may occur, especially in adolescence, further compromising self-esteem. Psychiatric sequelae, like depression and social anxiety, have been noted to occur. Together with the psychological sequelae, individuals would be concerned with the effect on cardiorespiratory function, chest pain, fatigue, frequent upper respiratory tract infections and growth.

The psychological sequelae become more prevalent in puberty and further development of self-awareness. The presence of the pectus malformations therefore would render the individual more self-conscious in terms of the anatomical difference. Therefore, together with physiological impairments, psychological and/or psychiatric manifestations need to be included as determinants of treatment.

Regarding quality of life, there are improvements in body image, sense of attractiveness, social interaction and overall satisfaction of the outcome after interventions.

In South Africa, telephonic interviews with individuals with pectus malformations were performed. The outcomes correlated with the literature with regards to psychological distress prior to the interventions. Marked psychological improvement was noted rendering these factors as important determinants for the consideration of treatment of pectus malformations.

SATS Paeds

Prenatal risk factors and environmental exposure associated with early childhood wheezing in the Mother and Child in the Environment (MACE) birth cohort

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Introduction. Prenatal and environmental exposure are contributing risk factors for development of a wheezing in early childhood.

Objective. To identify maternal and environmental risk factors associated with wheezing in children aged from birth to 24 months in the Mother and Child in the Environment (MACE) cohort.

Methods. The MACE cohort includes pregnant females selected from the public sector ante-natal clinics in Durban, South Africa ($n=1\ 140$), with similar socioeconomic profiles for inclusion in a longitudinal environmental study. Maternal health-related questionnaires were administered during pregnancy. The child was followed up clinically at 6, 12 and 24 months. Land use regression modelling was used to describe home address measures of exposure to nitrogen dioxide (NO₂).

Results. The mean (standard deviation) maternal age at delivery was 25.92 (5.96) years and the HIV infection rate was 35.6%. Children born from this cohort had a low-birthweight rate of 11.1%, a 7.3% preterm birth rate, a 70.3% exposed to passive smoking and 36.7% had a family history of asthma. Of the 466 children two years and younger that had at least one follow-up visit in the first 24 months of life, 109 (23%) had at least one incident of childhood wheeze as reported by a caregiver, with the majority of wheezy episodes ($n=63$) reported by the 12-month visit. Infants exposed to passive smoking (adjusted odds ratio (aOR) 1.737; 95% CI -1.073 - 2.811) and with a family history of asthma (aOR 2.724; 95% CI -1.650 - 4.496) were at an increased risk of wheezing. Ambient exposure to NO₂ was not associated with wheezing episodes in these infants (aOR 0.849; 95% CI -0.053 - 1.360).

Conclusion. Exposure to passive smoking and a family history of asthma were risk factors for wheezing in children from birth to 12 months. The absence of association of wheeze with NO₂ exposure could be related to a different pathogenic effect of the pollutant on lung parenchyma

Indoor pollution or tobacco smoke exposure and lung function at 3 years in an African birth cohort

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Introduction. Postnatal exposure to indoor air pollution (IAP) and environmental tobacco smoke (ETS) in early childhood has been associated with reduced lung function in childhood

Objective. To assess the impact of postnatal IAP and ETS on lung function and bronchodilator response (BDR) at 3 years.

Methods. Children enrolled in the Drakenstein Child Health Study, had lung function tested at 6 weeks and 3 years using standard and intra-breath measures of oscillometry. BDR was measured at 3 years. Indoor air pollutants (PM10, benzene) were measured in homes at 4 - 6 months postnatally. ETS was assessed by highest infant urine cotinine during 2 years.

Results. A total of 347 children were tested at 6 weeks and 3 years and BDR was successful in 224 (65%): 114 (50.7%) males, 110 (48.9%) were black Africans. The mean (standard deviation (SD)) BMI Z-score was 0.39 (1.2). The mean (SD) resistance (R10) and reactance at 10 Hz (X10) was 12.9 (3.4) hPa.L.s⁻¹ and -3.7 (2.1) hPa.L.s⁻¹, respectively. Post BD administration, the R10 was 11.0 (2.7) hPa.L.s⁻¹ and X10 was -2.8 (1.5) hPa.L.s⁻¹. Children exposed to high levels of ETS had higher R10 (R10 post: 3.0 hPa.L.s⁻¹; $p=0.01$; 95% confidence interval (CI) -1.4 - 4.5) and lower X10 (X10: -1.6 hPa.L.s⁻¹; $p=0.04$; 95% CI -3.0 - -0.1 and X10 post: -1.3; $p=0.03$; 95% CI -2.4 - -0.15) compared with unexposed children when adjusted for 6-week lung function. After BD, children exposed to high levels of PM10 had higher R10 (R10 at end-inspiration: 1.6 hPa.L.s⁻¹; $p=0.048$; 95% CI -0.2 - 3.1) and lower X10 (X10: -1.4 hPa.L.s⁻¹; $p=0.01$; 95% CI -2.4 - -0.3 and X10 at end inspiratory: -2.1 hPa.L.s⁻¹; $p=0.01$; 95% CI -3.4 - -0.8)

Conclusion. High exposure to ETS or IAP in early life is associated with impaired lung function at 3 years, adjusted for 6-week lung function. The effect of IAP was most notable after the use of a bronchodilator, highlighting lung development restriction.

Air pollution exposure and infant lung function in the MACE Cohort, South Africa

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Introduction. There is little known about the impact of air pollution exposure on infant lung function (ILF).

Objectives. This study aims to determine the relationship between antenatal exposure to oxides of nitrogen (NOx) and ILF outcomes at different time points, adjusting for maternal and infant risk factors.

Methods. Pregnant females (in the 'Mother and Child in the Environment' (MACE) birth cohort) were selected in the first trimester from public sector antenatal clinics in Durban, South Africa, and followed to delivery. NOx exposure was determined by land use regression modelling. ILF tests were performed at 1.5, 6, 12 and 24 months old in unsedated asleep children, following the ERS/ATS guidelines. Functional residual capacity (FRC) and lung clearance index (LCI) were measured by multiple breath washout using an ultrasonic flowmeter with 5% SF6 tracer gas. Measures of tidal breathing were obtained, with analysis of tests performed using W-breath software.

Results. Mean predicted antenatal NOx was 34.28 ug/m³ (13.75 - 42.24 ug/m³). Of the 263 ILF tests performed, 240 (91%) tests were accepted. In cross-sectional analyses, increasing NOx exposure resulted in a decline in FRC at 24 months, in LCI at 6 and 12 months, and a decline in tidal volume across ages. FRC was associated with a 0.6 mL (95% CI -2.12 - -0.72) decline with each unit increase in NOx. TV showed a 0.2 mL (95% CI -0.9 - 0.67) NOx-related decline, while LCI was not associated with exposure. For 62 participants with repeated measures, a consistent NOx-related decline was observed for all outcome measures.

Conclusion. ILF is associated with maternal NOx exposure during pregnancy and NOx exposure during early childhood. Repeated

measures at various time points provides a better understanding of the influence of NOx exposure on lung function and possibly lung development as the child ages.

Corrosive injury of the trachea in children

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Introduction. Ingestion of caustic substance is an uncommon medical presentation that has serious medical implications. A considerable amount of literature has been published on CSI injury to the gastrointestinal tract and the upper airway in children; however, limited information is available on tracheal injuries.

Methods. We report two cases of caustic ingestions who presented with respiratory symptoms requiring intubation, ventilation and subsequent multiple endoscopic operations to relieve their symptoms.

Results. In both these cases, there was a progression of the airway injury days to weeks after the injury. In the second case, the trachea had a normal appearance at the original bronchoscope and then progressed to stenosis. In this case, the contrast study did show gastroesophageal reflux, which may have contributed to the late onset of tracheal injury. The management of these cases is complicated. Surgery cannot be done early during the course of the disease because of the risk of ongoing fibrosis. The airway needs to be dilated with either rigid or balloon dilatation, until the fibrosing process has settled. The management may be complicated by the presence of oesophageal strictures as was the case in the second patient.

Conclusion. The ingestion of caustic substances still poses serious risks to children, the associated gastrointestinal injuries have been well studied with early and long-term complications well described; however, few cases of secondary respiratory injuries have been described. These may progress weeks to months after injury and repeat bronchoscopy is indicated. Ingestion of caustic fluid may cause severe tracheal injury or corrosion in children. Repeated airway dilatation may be lifesaving and until then, surgery can be done.

A rare clinical presentation of complicated childhood echinococcosis

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Introduction. We report a case of unusual childhood echinococcosis to raise awareness of the clinical presentation, varying features of childhood echinococcosis and the available, recommended modalities of investigating and managing complex echinococcosis in children.

Case. A 3-year-old boy presented tachypnoeic and hypoxic in room air, with a background history of a persistent cough for more than

2-months and low-grade fever. He had decreased ventilation on the right hemithorax, with scattered crepitations on both lung fields and a displaced apex; however, he was hemodynamically stable. His chest X-ray showed multiple oval opacities on both lungs. Contrast computed tomography (CT) scan of the chest and abdomen subsequently revealed multiple cystic lesions characteristic of hydatid cysts involving both lungs (largest on the right lower lobe), the liver, and the heart. Patient was managed as a complicated hydatid disease (ruptured cysts) with secondary pneumonic consolidation. He received anthelmintic (albendazole) treatment prior to surgery and postoperatively, some for 3 months and some for 6 months and followed up at monthly intervals. **Conclusion.** Hydatid disease is not uncommon in the paediatric age group. Though pulmonary and liver hydatid cysts are common in childhood, a high index of suspicion is required for hydatidosis presenting as the primary disease in unusual sites, especially in endemic areas.

Genome sequence of *Mycobacterium yongonense* RT 955-2015 isolated from a patient misdiagnosed with multidrug-resistant tuberculosis: First clinical detection in Tanzania

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Introduction. *Mycobacterium yongonense* is a recently described novel species belonging to the *M. avium* complex, which is the most prevalent aetiology of non-tuberculous mycobacteria associated with pulmonary infections, and poses diagnostic challenges in high-burden, resource-constrained settings.

Methods. Whole-genome shotgun sequencing and comparative microbial genomic analyses were used to characterise the isolate from a patient diagnosed with multidrug-resistant tuberculosis (MDR-TB) after relapse.

Results. Sequence analysis revealed that the RT 955-2015 strain had a high similarity to *M. yongonense* 05-1390(T) (98.74%) and *M. chimaera* DSM 44623(T) (98%). Its 16S rRNA showed similarity to *M. paraintracellulare* KCTC 290849(T) (100%), *M. intracellulare* ATCC 13950(T) (100%), *M. chimaera* DSM 44623(T) (99.9%), and *M. yongonense* 05-1390(T) (98%). The strain exhibited a substantially different rpoB sequence to that of *M. yongonense* 05-1390 (95.16%), but closely related to that of *M. chimaera* DSM 44623(T) (99.86%), *M. intracellulare* ATCC 13950(T), (99.53%), and *M. paraintracellulare* KCTC 290849(T) (99.53%).

Conclusion. In light of the OrthoANI algorithm and phylogenetic analysis, it was concluded that the isolate was *M. yongonense* of the Type

II genotype, which is an indication that the patient was misdiagnosed with TB/MDR-TB and received inappropriate treatment.

Real-time polymerase chain reaction v. conventional culture methods to detect respiratory pathogens in subjects with non-cystic fibrosis bronchiectasis

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Introduction. Non-cystic fibrosis (non-CF) bronchiectasis is increasingly recognised as a cause of chronic lung disease affecting children in developing countries. It is characterised by acute exacerbations, which results in repeated hospitalisation and lung function decline. Thus clinicians are interested in rapid and sensitive tests which are readily available and will assist in instituting the right treatment.

Objectives. The aim of this study was to compare polymerase chain reaction (PCR) with the conventional culture methods (CCM) to detect respiratory pathogens in subjects with non-CF bronchiectasis. Also important is the trade-off of cost increase from CCM to PCR v. the expected proposition of improved findings using PCR.

Methods. This was a cross-sectional study, using sputum taken from forty subjects who met the inclusion criteria and attended follow-up at Steve Biko Paediatric Pulmonology Clinic. Sputum was obtained through cough, cough-swab or sputum induction. The specimens were sent to the National Health Laboratory Service for sputum culture and a clinical medicine laboratory for respiratory multiplex PCR testing.

Results. Forty patients were enrolled. Sixty percent were male and the median age was 10 years. For PCR findings, 79% had a positive result and 21% had normal respiratory flora. These results were in contrast to CCM, which yielded 66% positive results and 34% normal flora. This indicates that PCR yielded more pathogens and the McNemar's test for symmetry was significant ($p=0.035$), with PCR detecting more positive results when CCM detects normal flora. The median turnaround time for PCR was significantly shorter than for culture (McNemar's test $p=0.001$; 2 v. 4 days). The actual cost of PCR is higher than CCM (ZAR1 333 v. ZAR247); however, faster turnaround times are generally associated with reduced hospital stay and reduction in overall cost.

Conclusion. There is a potential role for molecular techniques such as multiplex PCR to compensate for culture-based tests due to superior diagnostic yield and faster turnaround times. The reduced length of stay associated with early initiation of therapy and reduction of overall medical cost, mitigates the high costs of PCR.

Isotretinoin embryopathy: An unusual presentation of cough and noisy breathing

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Introduction. Isotretinoin embryopathy (fetal retinoid syndrome) is a characteristic pattern of mental and physical birth defects that result from maternal use of retinoids (oratane, roaccutane) in the first trimester of pregnancy. Isotretinoin is the most common agent used in the treatment of severe, recalcitrant, nodular-cystic acne refractory to conventional treatment

Methods. We present a case of an 11-month-old female patient who was delivered via elective caesarean section. The baby had good apgar scores at birth with no complications. On the 6 weeks follow up, mom reported that the baby had noisy breathing, persistent cough as well as episodes of choking during feeding. Physical examination revealed typical facial features of isotretinoin embryopathy: microphthalmia, retraction strabismus, high-arched palate, micrognathia, noisy breathing, and abnormal pinnae with pits. The patient was not in respiratory distress; however, there were coarse crackles bilaterally. The heart was normal. There was an umbilical hernia. Neurological examination revealed gross hypotonia, excessive head lag and developmental delay.

Results. The baby's clinical and radiological features were also reviewed in conjunction with the neurologist; the features were in keeping with embryonic disruption secondary to isotretinoin embryopathy. On further history, the mother confirmed that she was on oratane 10 mg oral twice a day prior to pregnancy. She was not aware of the teratogenic effects of oratane. She received counselling on the baby's current condition and follow-up. The baby was referred to an ENT specialist for hearing assessment, ophthalmology for visual assessment, speech therapy to assess the swallowing mechanism and aspiration. Further investigations for thymic abnormalities included serum calcium levels, chest x-ray, and CT chest. The patient was placed on omeprazole and further follow-up plans include growth monitoring and planning for feeding gastrostomy.

Conclusion. Isotretinoin embryopathy remains an important cause of teratogenicity; this case emphasises the importance of cautious treatment of acne in women of child-bearing age.

An unusual complication of paraffin ingestion: Pleural effusion

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Introduction. Paraffin poisoning is an important cause of preventable mortality in children. Paraffin is still a main source of fuel for cooking and lighting in poor household in South Africa. Infants are susceptible to accidental poisoning due to poor storage in cola bottles or in easily accessible areas. Paraffin poisoning is associated with chemical pneumonitis, hypoxia, bacterial pneumonia, pneumatocele, subcutaneous emphysema, rarely empyema and pleural effusion and death.

Case. We report a case of a 3 years 11 months old girl who presented to the emergency department with accidental ingestion of an unknown amount of paraffin. She was admitted with mild chemical pneumonitis. She was alert, had tachypnoea and hypoxia on room air. She responded well to oxygen at 2 L/min. All her other vital signs

were normal. Her initial chest X-ray was unremarkable, infective markers were normal (CRP 2). However, after 24 hours of admission, she had a worsening degree of respiratory distress and was intubated and ventilated. The arterial blood gas was normal. She was lethargic, hypoxic on 2 L/min and had pyrexia of 39°C. A repeat X-ray showed a left-sided pleural effusion. Repeat septic markers showed a rise in CRP to 305. Chest sonar confirmed a left-sided pleural effusion. Sonar-guided thoracocentesis did not yield any fluid, thus no sample was sent for analysis. The patient was started on paracetamol and amoxicillin-clavulanic acid. In light of the postulate that the pleural effusion was inflammatory in nature, prednisone 2 mg/kg/day was started as the pleural effusion was worsening. The patient required minimal ventilation settings after 72 hours and was extubated to nasal prongs oxygen. She completed five days of amoxicillin-clavulanic acid. She was weaned off oxygen after six days in the paediatric ward. She was discharged home and followed up after two-weeks. The repeat X-ray showed complete resolution of the pleural effusion. The patient was doing well clinically, with no residual respiratory distress and she had gained weight.

Conclusion. Paraffin poisoning is associated with pleural effusion, which is often haemorrhagic. However, there are limited data on the specific management of this condition.

A small girl with a big problem: Case report on giant mediastinal thymofibrolipoma

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Introduction. Thymolipoma occurs very rarely in children and account for less than 9% of thymic tumours which constitute less than 4% of mediastinal tumours in this age group. They have been associated with autoimmune diseases like myasthenia gravis. A rare histological variant includes thymofibrolipoma which consists predominantly of fibrous tissue.

We conducted a literature review and present a case report of a 12-year-old girl with progressive non-productive cough and shortness of breath. She was small-for-age and had orthopnoea, mild tachypnoea with Hoover's sign, a barrel-shaped chest and rested in the prone position.

Case. The chest radiograph showed bilateral opacification of the lung fields with total obscuration of the heart shadow and very small areas of apical lung markings. A CT scan demonstrated a giant mediastinal mass, extending anterior to both lungs. Mass effect on the pulmonary parenchyma was noted with areas of basal atelectasis and bronchiectasis. An initial tissue biopsy suggested a diagnosis of thymolipoma.

At median sternotomy, a giant anterior mediastinal mass was encountered extending into both thoracic cavities. Due to hemodynamic instability and hypercapnia intraoperatively, the fibrofatty mass was initially excised by dividing a 5 cm thick fibrous band over the heart. The mass was then systematically dissected from the superior extension by dividing the feeding vessels arising from the superior intercostal artery and the internal mammary artery and the draining vessels to the brachiocephalic vein. A clear capsular plane of dissection was not achieved over the lungs and phrenic nerves due to

thick fibrous adhesions over the pulmonary vasculature, visceral pleura and mediastinum. The mass consisted of fatty lobules with areas of thick fibrotic changes and weighed 721 g. Histology showed extensive dense fibrosis and the mature adipose tissue had a fibrous capsule and intimately associated with unremarkable thymic tissue containing cortex, medulla and Hassall's corpuscles. These features are consistent with thymofibrolipoma.

Conclusion. Thymolipomas can proliferate to massive dimensions before significant symptoms develop. The potential complications to the respiratory system and technically challenging surgery must not be underestimated. Sternotomy provides optimal exposure and may require a clam shell extension. Minimally invasive resections have been reported.

A rare cause of diffuse pulmonary nodules in a child

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Introduction. An 11-year-old HIV-uninfected male was referred to our centre with probable miliary tuberculosis (TB) that was failing to respond to treatment after 2 months of 4 drug therapy. He had presented originally with a 2-month history of constitutional TB symptoms (unintentional weight loss, difficulty breathing and occasional night sweats). A chest X-ray performed at the time revealed a diffuse nodular pattern consistent with miliary TB. There was no family history of TB. He was cachexic, had difficulty breathing and a productive cough, without haemoptysis. He had generalised shotty lymphadenopathy with cervical nodes more prominent. He was not clubbed but he was hypoxic. Chest auscultation revealed diffuse fine crackles.

Case. This is a case report of a patient seen as an inpatient, who presented with diffuse nodular pulmonary infiltrates. A chest X-ray revealed extensive diffuse nodular pulmonary infiltrates. A laboratory panel of investigation was initiated and excluded common causes of diffuse parenchymal lung disease, including infections (sputum and nasopharyngeal aspirate tests excluded viral, mycoplasma, fungal and mycobacterial causes), while haematological testing excluded autoimmune disease, HIV and a PID. Bronchoscopy and bronchoalveolar lavage (BAL) revealed no pathology. A CT chest revealed thyroid nodules and diffuse pulmonary nodules consistent with metastatic lesions from a thyroid carcinoma.

Excisional biopsy of the cervical lymph nodes confirmed a follicular variant of papillary thyroid carcinoma. The patient had a total thyroidectomy followed by radiotherapy ablation therapy. He remains alive.

Conclusion. Paediatric thyroid carcinoma (PTC) is very rare, estimated to be around 3% of all paediatric malignancies. Of all PTC, papillary thyroid carcinoma is the most common, accounting for 83%. Although PTC is rare as an entity, the presence of thyroid nodules in children is most often associated with malignancy compared to

thyroid nodules in adults which will most likely be benign. Papillary thyroid carcinoma presents as painless, subtle, symptomless thyroid nodules or cervical nodes that are often incidentally found by parents or health care workers. It can also present with a miliary pattern like pulmonary infiltrates (as in our patient), even then patients often remain asymptomatic without notable pulmonary symptoms. Treatment consensus is that for metastatic disease, a total thyroidectomy followed by radioactive iodine treatment is gold standard treatment for such patients. The prognosis is generally good for PTC, especially in those patients that are diagnosed early. Rare pulmonary diseases should be considered in children not responding to conventional therapy.

Mediastinal fibromatosis in a child that presented with a superior vena caval syndrome

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Introduction. An 8-year-old HIV-uninfected male patient, who was previously treated for pulmonary TB in 2015, presented with a non-productive cough for 2 weeks. This was associated with weight loss and neck and facial swelling. He was not clubbed nor was there generalised lymphadenopathy. He had distended neck veins but no other signs of cardiovascular involvement. He was not in respiratory distress and chest examination yielded normal findings.

Case. A case report of a child that presented with a superior vena cava syndrome who was later diagnosed with a mediastinal mass. A chest radiograph revealed a right-sided anterior mediastinal mass. All haematological testing was normal. TB was excluded with appropriate testing. A computed tomography (CT) chest scan confirmed a right-sided anterior mediastinal mass with mass effect on the adjacent structures and compression of the superior vena cava (SVC) and a CT-guided biopsy of the mass suggested a desmoid type fibromatosis. Complete resection of tumour was done and a diagnosis of fibromatosis was confirmed histologically. The patient responded well postoperatively and the SVC syndrome resolved. No chemotherapy or radiotherapy was required.

Conclusion. Fibromatosis is a very rare, slow-growing, non-metastatic proliferation of fibrous tissue associated with excessive collagen formation. It is classified as superficial or deep fibromatosis. Deep fibromatosis is often termed a desmoid tumour. Desmoid tumours, although not malignant, have a tendency to be locally aggressive as they tend to adhere to and also exert mass effects on local and surrounding structures. Management of deep fibromatosis includes complete resection of the mass with wide margins. However; this is not always possible as these tumours tend to tenaciously adhere to other structures. Radiotherapy and chemotherapy have also been employed especially for incompletely resected and for recurring tumours. In the recent years, 5 cases of mediastinal fibromatosis in children have been diagnosed. Though previously a very rare

cause of a mediastinal mass, fibromatosis is now emerging as a not so uncommon occurrence. Therefore, mediastinal fibromatosis should now be considered an important differential diagnosis in the evaluation of anterior mediastinal masses in children.

The South African Cystic Fibrosis Registry Initiative (SACFRI): Implementation challenges and initial data

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Introduction. Cystic Fibrosis (CF) occurs with varying incidence throughout the world. Improved survival and care are partly attributed to knowledge gained from international CF registries. A multi-centre public-private CF registry initiative was launched in South Africa (SA) in April 2018.

Objectives. The aim of this report is to describe implementation challenges and initial data collection.

Methods. CF diagnosis criteria and annual data collection variables were adopted from the European CF registry. Institutional research ethics approval was obtained for participating sites/private practices with informed consent/assent a requirement for participant inclusion. Demographic profile, diagnosis information and cross-sectional lung function data from 1 January to 31 December 2017 is described.

Results. Obtaining several research ethics approvals and informed consent, incomplete medical records, unconfirmed ($n=35$, 11%) CF diagnosis information, infrequent follow-up visits and fragmented care were obstacles encountered. By April 2019, 329 people ($n=179$; 54.4% females) with CF representing Western Cape, Eastern Cape, KwaZulu-Natal and private clinics in Gauteng were entered into the registry with the following age distribution: 58 (18%) <6 years, 167 (62%) 6 - 18 years and 104 (38%) >18 years. Most were white ($n=227$; 69%) or mixed/other race ($n=82$; 27%); a minority black African ($n=15$, 4.6%). The median age at diagnosis was 6 months (IQR 0.1 - 3 years) with 50 (15%) presenting with meconium ileus. At least one sweat test was documented in 201 (61%). Reported frequency of CFTR mutations was: p.Phe508del 60%; other 15%; unknown 11% and c.2988+1G>A 7%. Annual review data of 306 people was available for 2017: 150 (49%), 97 (32%) and 59 (19%) in private, public or private/public sectors respectively. Chronic *Pseudomonas aeruginosa* infection was documented in 97 (32%) however data on *P. aeruginosa* infection status was missing in 61 (20%). Best forced expiratory volume in 1 second (FEV_1) z-scores recorded for ages ≥ 6 years ($n=213$) was -1 in 133 (62%) and -3 in 62 (29%).

Conclusion. Despite challenges and incomplete data collection, the SACFRI has shown initial epidemiological data of CF patients in SA. Representation from all provinces and intensified data collection strategies are needed.

SATS Adult

Cigarette smoke exposure augments the expression of pneumococcal antibiotic resistance genes

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Introduction. *Streptococcus pneumoniae* is a leading cause of morbidity and mortality worldwide, with emerging antimicrobial resistance of considerable concern. While smoking is a well-recognised risk factor for severe invasive pneumococcal disease, little is known about possible effects of cigarette smoke exposure on pneumococcal virulence mechanisms, and especially the expression of antibiotic resistance. Importantly, smoking has been linked, among other factors, to bacterial mutagenesis.

Objective. The aim of the present study was to examine the effects of cigarette smoke condensate (CSC), alone and in combination, with the macrolide, clarithromycin, on the expression of the macrolide resistance genes, *mef(a)* and *erm(b)*, in strains 521 and 2507 of the pneumococcus, respectively, as well as that of the ABC-transporter gene, *sp2003*, which may also be involved in the development of antibiotic resistance.

Methods. Following exposure of the *S. pneumoniae* strains 172 (macrolide-susceptible, control strain), 521 (MIC 2 mg/L) and 2507 (MIC >256 mg/L) to CSC (80 and 160 mg/L) and clarithromycin (0.125 mg/L, 2 mg/L and 8 mg/L, respectively), individually and in combination, the RNA was extracted, cDNA synthesised and real-time qPCR performed, using standard techniques. Data were analysed by comparing the relative change in expression of the target genes to that of the reference genes and is expressed as fold increase.

Results. As expected, exposure of both test strains of the pneumococcus to clarithromycin resulted in substantial upregulation of both macrolide resistance genes, which was significantly ($p<0.001$) augmented by prior exposure to CSC in the case of *erm(b)*, but not *mef(a)*. Somewhat unexpectedly, exposure of strain 2507 to CSC (160 mg/L) alone (in the absence of clarithromycin) resulted in significant ($p<0.05$) expression of the *erm(b)* gene. In addition, the ABC-transporter gene was upregulated in all three strains, particularly following exposure to both clarithromycin and CSC.

Conclusions. Exposure of the pneumococcus to CSC prior to treatment with clarithromycin resulted in increased expression of all three resistance genes, suggesting that smoking could favour the selection of macrolide resistance, as well as other antibiotic resistance mechanisms, in this dangerous pathogen. The findings of this study shed new light on the link between smoking and increased resistance gene expression by the pneumococcus.

In vivo and in vitro studies to investigate the role of autophagy in human tuberculosis

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Introduction. *Mycobacterium tuberculosis* (*M. tb*) is one of the world's most successful human pathogens infecting ~2 billion people worldwide. Although there are effective chemotherapies for TB, the disease remains out of control due to prolonged and toxic treatment. Shorter regimens are urgently required to control TB. Drug resistance TB also threatens to derail TB control, these unmet needs could be addressed by the identification and development of host directed therapeutic agents for TB. Manipulation of innate immune response, including autophagy, may lead to the identification of cellular pathways that could be exploited to develop host directed TB therapeutic agents. Whether the induction of autophagy by metformin (met) and nitazoxanide can promote *M. tb* stasis remains unclear.

Methods. Blood and/or bronchoalveolar lavage (BAL) fluid were obtained from participants with varying degrees of *M. tb* susceptibility, presumed latent TB infection, presumed sterilising immunity, previous TB, and recurrent TB. Expression of LC3II protein, the marker of autophagy from peripheral blood monocyte cells, monocyte-derived macrophages and BAL cells were quantified by western blot and confocal microscopy. Cell cultures were treated *in vitro* with met, nitazoxanide and starvation media in the presence or absence of bafilomycin. Some of the participants received bronchoscopic-instilled BCG with follow-up bronchoscopy at day 3; autophagic proteins (LC3II) were measured pre- and post-treatment in the presence and absence of bafilomycin.

Results. Met and starvation was a potent inducer of autophagy in BAL cells ($p=0.0008$ compared with no met $n=22$) and for starvation ($p=0.0004$ compared with control $n=19$). *In vivo*, BCG was an inducer of autophagy ($p=0.0005$ compared with saline control $n=13$).

Conclusion. These preliminary results demonstrate that met, starvation and BCG induced autophagy in alveolar cells. These findings have implications for new host-directed therapies. The next step is to determine if the relevant pathways influence mycobacterial survival *in vitro*.

Characterisation of adult asthmatic patients followed up at pulmonology outpatient clinics in Luanda, Angola

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Introduction. Asthma is one of the most common chronic diseases, affects all ages, and is one of the most frequent causes of visits to emergency services around the world. One of the objectives of the Global Alliance against Chronic Respiratory Diseases (GARD) is to monitor these diseases and their determinants. However, studies on features of asthma in adults in Africa are sparse and in Angola there is none.

Objective. To evaluate clinical features of asthma and factors associated with its control.

Methods. A cross-sectional study was conducted from April 2018 to March 2019 in patients who were ≥18 years old, with clinically confirmed asthma and who were followed up at pulmonology outpatient clinics at the military hospital in Luanda. Asthma was assessed in accordance with GINA (global initiative for asthma) criteria, lung function (spirometry) was performed and analysed in accordance with ATS/ERS criteria, and sensitisation to aeroallergens was determined by skin prick test (SPT) positivity. Asthmatic patients with previous pulmonary tuberculosis, or chronic obstructive pulmonary disease (COPD) were excluded.

Results. The sample consisted of 305 asthmatic patients (mean age 41.3, median 41.0 (18 - 86 years)), 56.1% female. Of these, 6.9% had intermittent, 62.0% mild persistent, 26.9% moderate and 4.3% severe asthma. In 56.1% of cases, asthma was associated with allergic rhinitis. Regarding asthma control, 28.2% patients had controlled asthma, 36.4% partially controlled and 35.4% uncontrolled. Only 39.0% of patients used controller medication but irregularly, and 25.9% only used rescue medication. Around 30% of patients had more than 5 exacerbations in the previous year and 44.6% had taken oral corticosteroids. Spirometry was normal in 21.6% of patients, showed mild obstruction in 47.9% and moderate to very severe obstruction in 30.5% of patients. Most patients (66.9%) were sensitised to aeroallergens, most frequently dust mites, cat and dog epithelia, and fungi.

Conclusion. Most adult asthmatic patients in Luanda have only partially controlled or uncontrolled symptoms, are not on or irregularly use inhaled controller medication, have concurrent allergic rhinitis and are sensitised to aeroallergens.

'I am a new person' – the benefits of a pulmonary rehabilitation programme with limited resources: A case study

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Introduction. There is unequivocal evidence that pulmonary rehabilitation (PR) is beneficial in the management of chronic obstructive pulmonary disease (COPD). Despite this, there is limited availability of PR in South Africa. A PR service was established at a tertiary hospital with the aims of service provision and to establish a

programme which can be delivered with minimal resources and have sustained benefits.

Methods. The 12-week programme consisted of individually tailored, weekly supervised exercise and education sessions, as well as a home programme. Prior to starting the programme, medications were optimised and comprehensive assessment was conducted. An educational booklet and exercise diary were provided. Education focused on nutrition and breathing techniques and exercise focused on strengthening large muscle groups and endurance. All exercises were progressed according to performance. Assessment was repeated at the end of the programme, 3 months, 9 months and 12 months.

Results. A 76-year-old man with GOLD stage 4 COPD completed the 12-week PR programme. He is an ex-smoker with a ~40 pack-year smoking history. Co-morbidities include hypertension, diabetes mellitus and a previous stroke (right hemiplegia). In the 6 months preceding PR, he had three exacerbations which required medical attention. He required a caregiver at home to assist in daily activities. Following the programme, clinically significant and sustained improvements in the cycle endurance test (baseline 5:55 min; 3 months 11:00 min; 9 months 8:23 min) and 6-minute walk test (baseline 270 m; 3 months 372m; 12 months 383m) were found despite a decline of 11% in fev1 over the year. Marked improvement in his quality of life scores (SGRQ score baseline 71.61; 3 months 23.39; 12 months 31.19) were found. He expressed great satisfaction with the programme: 'I am a new person', 'I could play dominoes the whole day and not think about breathing'. For a year following the programme, he has not had to seek healthcare for respiratory problems and has not needed a caregiver.

Conclusion. Pulmonary rehabilitation is possible with minimal resources and can have significant lasting benefits to patients.

Serum chitotriosidase activity in patients with sarcoidosis and pulmonary tuberculosis

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Introduction. Human chitotriosidase is a chitinase enzyme selectively expressed by activated macrophages. It was initially described as having markedly elevated activity in Gaucher's disease patients. An increase in chitotriosidase activity has been described previously, both in serum and in bronchoalveolar lavage, of patients with sarcoidosis. Chitotriosidase may be used as a potential biomarker for the diagnosis and monitoring, and as a prognostic marker in patients with sarcoidosis.

Objective. To analyse serum chitotriosidase activity in a South African cohort of patients with sarcoidosis to verify the reported increase with respect to controls. We also compared serum chitotriosidase levels

in patients with sarcoidosis and tuberculosis, two granulomatous disorders of different aetiology, as South Africa is a high-burden area for tuberculosis.

Methods. Chitotriosidase activity was assayed in serum of 12 biopsy-proven sarcoidosis patients before treatment, 9 sarcoidosis patients after treatment, 10 confirmed pulmonary tuberculosis patients, prior to treatment and 12 healthy controls. Plasma chitotriosidase was assayed by a previously described method using 4-methylumbelliferyl- β -D-N,N',N''-triacetylchitotriose as a substrate.

Results. We found significantly higher serum chitotriosidase activity in sarcoidosis patients than controls ($p=0.05$), both in untreated and treated patients. Sarcoidosis patients had higher chitotriosidase levels than tuberculosis patients; however, this did not reach statistical significance. While in patients with tuberculosis chitotriosidase activity was found to be lower than in sarcoidosis patients, it was higher than in controls.

Conclusion. To the best of our knowledge, this is the first report of chitotriosidase activity analysis in the serum of patients with sarcoidosis and pulmonary tuberculosis in South Africa. Although the mechanisms leading to the increase in chitotriosidase activity in sarcoidosis is still unknown, this enzyme may be specifically involved in the pathogenesis of the disease. Further studies with greater number of patients are required to confirm these results. Determining whether chitotriosidase could be a marker with diagnostic or prognostic value in sarcoidosis remains to be established.

Clinical features of patients with sarcoidosis in Johannesburg, South Africa

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Introduction. Sarcoidosis is a multisystem granulomatous disease of unknown aetiology most commonly affecting the lungs. Because of its similarities, both clinically and radiologically, and particularly in a high tuberculosis (TB) prevalence region, sarcoidosis is frequently misdiagnosed as TB.

Objective. To study the profile of sarcoidosis in South African patients in Gauteng Province.

Methods. Retrospective study of patients attending the Charlotte Maxeke Johannesburg Academic Hospital pulmonology services over a 5-year period.

Results. Of 102 patients with sarcoidosis, 59 were black, 28 Indian, 8 white and 7 coloured (mixed race). There were 69 females (68%) and 33 (32%) males. The majority (87%) were non-smokers. The median age of the group was 44.6 years. Seventeen percent had been treated for tuberculosis prior to being diagnosed with sarcoidosis. Two patients developed active TB while being treated with corticosteroids for sarcoidosis. Four patients contracted human immunodeficiency virus infection. The most frequent chronic comorbid illnesses were

hypertension (32%), obesity (18%), diabetes mellitus (16%), heart failure (16%), asthma (12%), and gastro-oesophageal reflux disease (12%). Cough was the most common presenting symptom in 82%, dyspnoea (52%), skin lesions other than erythema nodosum (33%) and crackles (32%) were the other salient clinical manifestations. The majority (48%) had stage II chest radiographic changes. The skin (28%), mediastinal lymph nodes (26%) and transbronchial lung biopsies (26%) were the most frequent sites confirming granulomatous inflammation. Twenty percent had obstructive airways disease on lung function testing. Systemic corticosteroids were indicated in 84% of patients.

Conclusion. Sarcoidosis is often misdiagnosed as TB in South Africa. The most frequent sites for histological confirmation were skin, mediastinal lymph node and transbronchial lung biopsy. Comorbid illnesses were common and should be screened for and treated. The majority of patients in this cohort with sarcoidosis had an indication to be treated with corticosteroids.

Spectrum of diffuse parenchymal lung disease with special reference to idiopathic pulmonary fibrosis: experience at Charlotte Maxeke Johannesburg Academic Hospital

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Introduction. Diffuse parenchymal lung diseases (DPLD) encompass a group of diseases with a wide range of causes with varied presentations and prognosis. The known causes include occupational or environmental exposure, drug-induced lung diseases, hypersensitivity pneumonitis and connective tissue disease (CTD). Among the DPLD with unknown cause, idiopathic pulmonary fibrosis (IPF) is the most common and has the worst outcome. The earlier publications from this country described cryptogenic fibrosing alveolitis. With subsequent characterisation of idiopathic interstitial pneumonia (IIP), and an increase in the burden of IPF reported worldwide, we evaluated the clinical features of patients with IPF in the South African context.

Objective. To evaluate the clinical spectrum of DPLD encountered in Johannesburg, South Africa, and to describe the clinical profile of patients with IPF.

Methods. A retrospective record review of patients who attended the Charlotte Maxeke Johannesburg Academic Hospital (CMJAH) respiratory clinic in the 5-year period from January 2011 to December 2015 was undertaken. Patients with DPLD were identified and the specific diagnoses were noted. The records of patients with IPF were further analysed.

Results. We identified 132 patients with DPLD. Sarcoidosis (37.8%), IPF (21.2%), connective tissue disease-associated diffuse parenchymal lung disease (CTD-DPLD) (14.3%), and hypersensitivity pneumonitis (HP) (9.8%) were the four most common types. IPF was seen in all racial groups. Of the 28 patients with IPF in our cohort, there was

a slight female predominance (1.3:1). The mean age of the patients in our study was 63.8 years and the majority were white. Cough (96.4%), dyspnoea (92.8%) and bilateral crackles (96.4%) were the most common clinical features. The majority of patients (78.5%) were diagnosed by high-resolution computerised tomography (HRCT) scan.

Conclusion. IPF is the second most common DPLD disease encountered after sarcoidosis at CMJAH. IPF is seen in all racial groups in Johannesburg, South Africa, and the characteristics of patients with IPF are similar to those seen in other parts of the world.

Altered serum miRNA expression in patients with sarcoidosis – a pilot study

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Introduction. Sarcoidosis is a granulomatous disease of unknown aetiology. The disease has an important inflammatory and immune component; however, the immunopathogenesis is not completely understood. Micro ribonucleic acids (microRNAs or miRNAs) are groups of small, single-stranded, non-coding RNAs. Their major identified function involves mediating the posttranscriptional silencing of target genes.

Recently, the role of miRNAs has attracted attention as both being involved in the pathogenesis, and serving as disease markers, in numerous diseases. Not much is known about the role of miRNAs in sarcoidosis and is being unravelled by many workers.

Objective. To investigate the serum expression of ~800 miRNAs in patients with sarcoidosis ($n=6$) and race-, age- and gender-matched healthy controls.

Methods. Whole blood samples were collected in EDTA tubes, processed and the plasma retained. RNA was extracted from the stored plasma samples using the Qiagen miRNeasy mini kit and concentrated using a salt-ethanol precipitation. The RNA extracted was analysed using an nCounter miRNA human V3 expression assay. Twelve samples (6 patient and 6 matched controls) were analysed. Data were analysed using the nSolver analysis software.

Results. One pair of results was excluded as a result of cellular RNA contamination. Therefore, 5 patient and 5 matched control samples were analysed. After excluding miRNAs that were below background levels, 145 miRNAs could be analysed. On applying a Bonferroni correction, the only miRNA that was significantly different was miRNA let-7a-5p which was significantly overexpressed in patients compared with controls (patient count of 218.39 and control count of 1.54 (141.69-fold change; $p=0.0003$). Of the 145 miRNAs, many were differentially expressed in patients compared with controls, although not reaching statistical significance.

Conclusion. To the best of our knowledge, this is the first miRNA report of differentially expressed miRNAs in the serum of patients with sarcoidosis and matched healthy controls in South Africa. The results

obtained suggest that miRNAs may have a role in the pathogenesis of sarcoidosis. Further studies are needed to determine whether these molecules have diagnostic or prognostic implications.

Gestational lung cancer: A case report and brief literature review

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Objective. To share ethical dilemmas of a patient with lung cancer falling pregnant.

Case. A 40-year-old female patient (P1G2), was referred to the respiratory service for assistance with care. She had been diagnosed with a poorly differentiated and advanced (stage IV) squamous cell carcinoma of the left lung two years prior. Her disease was poorly responsive to palliative chemotherapy and had continued to progress clinically and radiologically, despite 6 cycles of paclitaxel and cisplatin. At follow-up, 9 months since her last chemotherapy, she was found to be 20 weeks pregnant.

Conclusion. Outcomes, as in the non-pregnant states, are determined by the disease stage; with early stage disease associated with longer term survival. Lung cancer can adversely affect pregnancy. However, most pregnant mothers with lung cancer give birth to healthy babies. Pregnancy can potentially delay the management of lung cancer; risking tumour progression. Radiotherapy with abdominal shielding can be offered safely. Surgery is usually delayed until after the first trimester. Chemotherapy during the first trimester is discouraged. When started in the 2nd trimester, neonatal outcomes have been good. Publications on platinum-based and targeted chemotherapy are meagre. Extensive counselling is important.

Case report: Broncho-alveolar carcinoma presenting as unusual non-resolving cavitating pneumonia

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Introduction. We report a case of an unusual radiological pattern of broncho-alveolar carcinoma (BAC) presenting as non-resolving cavitating pneumonia. BAC (mucinous adenocarcinoma) typically presents as a solitary lesion, consolidation or diffuse radiological pattern. A 50-year-old female (non-smoker) presented with more than 1 year history of a cough with mucopurulent sputum consistent with bronchorrhea, episodes of hemoptysis and significant loss of weight. Her physical examination revealed temporal wasting and bilateral crackles. A chest X-ray and CT chest done in 2017 revealed a cavitating lesion in the medial basal segment of the right lower lobe. In keeping with the radiological pattern, a work up for infective process was done-sputum for Gene-Xpert, culture, cytology and blood tests (full blood count, kidney function and autoimmune studies) were all negative. She subsequently received multiple

courses of antibiotics as she was being treated for a cavitating pneumonia but with no symptomatic relief. A follow-up CT chest in 2018 revealed cavitating lesion with surrounding ground glass opacities. In 2019, as a result of her clinical deterioration, CT chest showed multiple cavitating lesions in both lung fields. An open lung biopsy was consistent with invasive mucinous adenocarcinoma with a negative epithelial growth factor receptor (EGFR). She subsequently progressed to respiratory failure and demised prior to treatment initiation.

Case. Written consent was obtained from the family. This case highlights an unusual radiographic presentation of BAC, as non-resolving cavitating pneumonia. BAC is a variant of NSCLC, accounting for 6.5% of lung malignancies. It is commonly described in younger females between 40 and 70 years of age, non-smokers with common symptomatology of cough, shortness of breath, haemoptysis and fever. Bronchorrhea is a rare and late manifestation. It has varying radiographic patterns including solitary lesion, consolidation and a multifocal or diffuse pattern. Mucinous form of adenocarcinoma is the most aggressive form that responds poorly to treatment.

Conclusion. BAC should be considered in the differential diagnosis of patients who present with cavitating lesion on chest radiograph as delay in diagnosis would lead to poor outcome as seen in our case.

A very short series of uniportal pulmonary resection

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Introduction. We present our evolving experience of uniportal video-assisted thoracoscopic surgery (VATS) pulmonary resection, starting January 2018 until March 2019, where 14 patients were operated.

Objective. To evaluate safety, surgical difficulties and outcomes in our spectrum of disease.

Methods. All patients who presented for pulmonary resection were included, making for 8 inflammatory lung disease, 1 repeat surgery for post tuberculous bronchiectasis with haemoptysis, 4 bronchial carcinoma and 1 lymphoma patients; of whom one had planned single coronary artery and aortic valve surgery a week prior pulmonary resection. Operative outcomes were evaluated, for need to convert to thoracotomy, bleeding, reasons for intercostal drain stay, operative complications and mortality.

Results. Ten patients were operated on without conversion, 4 patients were converted for intraoperative bleeding from pulmonary artery injury these were among the first 6 patients. The rest of the patients had shorter ICU stays and less pain complaints. While the periods and reasons for drain removal were similar to the thoracotomy patients, there were no records of post thoracotomy-like pain and bleeding was not a concern. There was one mortality in a converted patient on day 14.

Conclusion. Uniportal VATS pulmonary resection is an alternative and offers advantage to thoracotomy. Hilar access is important, reduce

bleeding complications. Inflammatory lung disease offers difficulty, from hard adherent lymph nodes and fibrosis. This challenge is an opportunity for patient and meticulous dissection for the benefit such patients. An additional port may be necessary in severe fibrotic disease.

Old foe – empyema thoracis; uniportal decortication – new weapon

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Introduction. We present our evolving experience of uniportal video-assisted thoracoscopic surgery (VATS) empyema thoracis intervention, starting September 2016 till March 2019, where 91 patients were operated. This is an observational study of patients who required decortication in a private practice setting.

Methods. All patients who presented for decortication were included, making for 2 bronchial carcinoma pleural disease; 50 tuberculous; 1 mesothelioma, 1 repeat surgery in a previous thoracotomy patient and 37 non-specific empyema thoracis patients were subjected to this surgery. Operative outcomes were evaluated for the need to convert to thoracotomy, bleeding, reasons for intercostal drain stay, persistence of air leak, operative complications and mortality.

Results. A total of 88 patients were operated on without conversion; 3 were converted for intraoperative bleeding and difficult plane of decortication and there were 3 empyemectomies. The remaining patients had shorter ICU stays, less pain complaints, length of time of drains inserted and reasons for drain removal were similar to the thoracotomy patients, no records of post thoracotomy pain and 1 postoperative bleeding which led to the only surgical mortality.

Conclusion. Uniportal VATS decortication is a surgical alternative to thoracotomy, offering advantages. Pleural panoramic access is important, reduce diaphragm injury, bleeding complications and surgical demand on the patients. Visceral pleural injury remains a concern with resultant air leak, though currently not showing difference from the thoracotomy patients. The current instruments are inadequate when severe organised disease is encountered. Better and adaptable instrumentation needs to be developed.

Autologous blood patch pleurodesis for the management of persistent air leak in secondary spontaneous pneumothoraces

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Introduction. Persistent air leak (PAL) may complicate the course of secondary spontaneous pneumothoraces and presents a particular challenge in patients who are deemed unfit for any form of surgical intervention. Although autologous blood patch pleurodesis (ABPP) has been used by thoracic surgeons for decades to manage PAL following pulmonary surgery, its role in the management of spontaneous pneumothoraces remains less well defined.

Objective. The aim of this prospective observational study was to assess the feasibility of ABPP in our setting.

Methods. Thirty-eight consecutive patients who had secondary spontaneous pneumothoraces with confirmed PAL and who were deemed unfit for surgery were recruited. The most common underlying causes were post-TB bronchiectasis ($n=22$) and COPD ($n=12$). A total of 80 - 120 mL of the patient's own unheparinised blood was injected via a 50 mL bladder syringe into the pleural cavity via an intercostal drain (ICD). The ICD was clamped for one hour, followed by chest radiography and if the air leak ceased, the ICD was removed. ABPP could be repeated once within 48 hours if PAL was still present, and was deemed successful if the ICD could be removed within 72 hours of the final procedure.

Results. ABPP was successful in 31 (81.6%) subjects and 15 (39.5%) required two procedures. The mean (standard deviation) duration of PAL prior to ABPP was 15 (5) days and the mean time to removal of the ICD was 1.6 days after the final procedure. The ICD could be removed within 24 hours in 17 (44.7%) subjects. Only one procedure-related complication (haemoptysis) was observed.

Conclusion. ABPP appears to be a feasible, safe and inexpensive intervention for PAL in poor surgical candidates with spontaneous pneumothoraces.