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Diffuse Idiopathic Skeletal Hyperostosis (DISH)

Case of the month - November

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Queensland Government

Clinical background

Diffuse Idiopathic Skeletal Hyperostosis (DISH), or Forestier disease, describes a disease process that involves proliferation of bony growths and ossification of tendons and ligaments at sites where they attach to the skeleton (entheses)[1]. These ossifications our commonly found in the spinal column, classically described as 'flowing ossifications along the anterolateral aspect in at least three successive vertebral levels or four contiguous vertebrae'[2]. The disease process is thought to develop between the ages of 30-50 but is rarely reported in patients under 50[3]. The prevalence of DISH tends to increase with age to around 14% of the population and is around three times more likely to occur in men[4]. Clinically, DISH can have no symptoms and is often noted as an incidental finding on imaging[5]. However, the classic presentation of DISH is that of an older patient with increasing back pain and limited range of spinal motion, which is often accompanied by an increased susceptibility to unstable spinal fractures[1]. As there is typically a baseline level of spinal pain, there may be a delay in diagnosis putting the patient at risk of spinal cord injury or further trauma[6].

Patients with DISH may also present with neurological symptoms such as dysphagia, aspiration pneumonia, sleep apnoea and hoarseness depending on the location of the ossifications and if they impinge on the nerves leaving the spinal column[2,7]. The disease process has also been linked to metabolic disorders such as diabetes mellitus, gout and hyperlipidaemia[2], though the link between the two is not entirely clear as the pathophysiology of DISH is yet to be fully understood[1]. There is no cure for DISH, though the symptoms can be managed and relieved with physiotherapy and analgesia. Surgical intervention may be considered in certain severe cases where the ossifications on the cervical spine compress the spinal cord, to relieve the impingement on the affected nerves and maintain quality of life[8].

With regards to lung function and its relationship with DISH, there has been links drawn between restrictive spirometry and lung volumes and the disease process[3,9], as well as with respiratory failure[5]. The mechanical limitations on both the spine and the ribcage resulting from the ossification process lend itself to a restriction-like pattern, stopping the thorax from expanding to its full extent. The studies performed reference COPD study datasets and are thus focused on current and former smokers[3,9]. A possibility of mechanical airway obstruction has also been suggested as a result of bone changes in the anterior cervical spine[7,10].

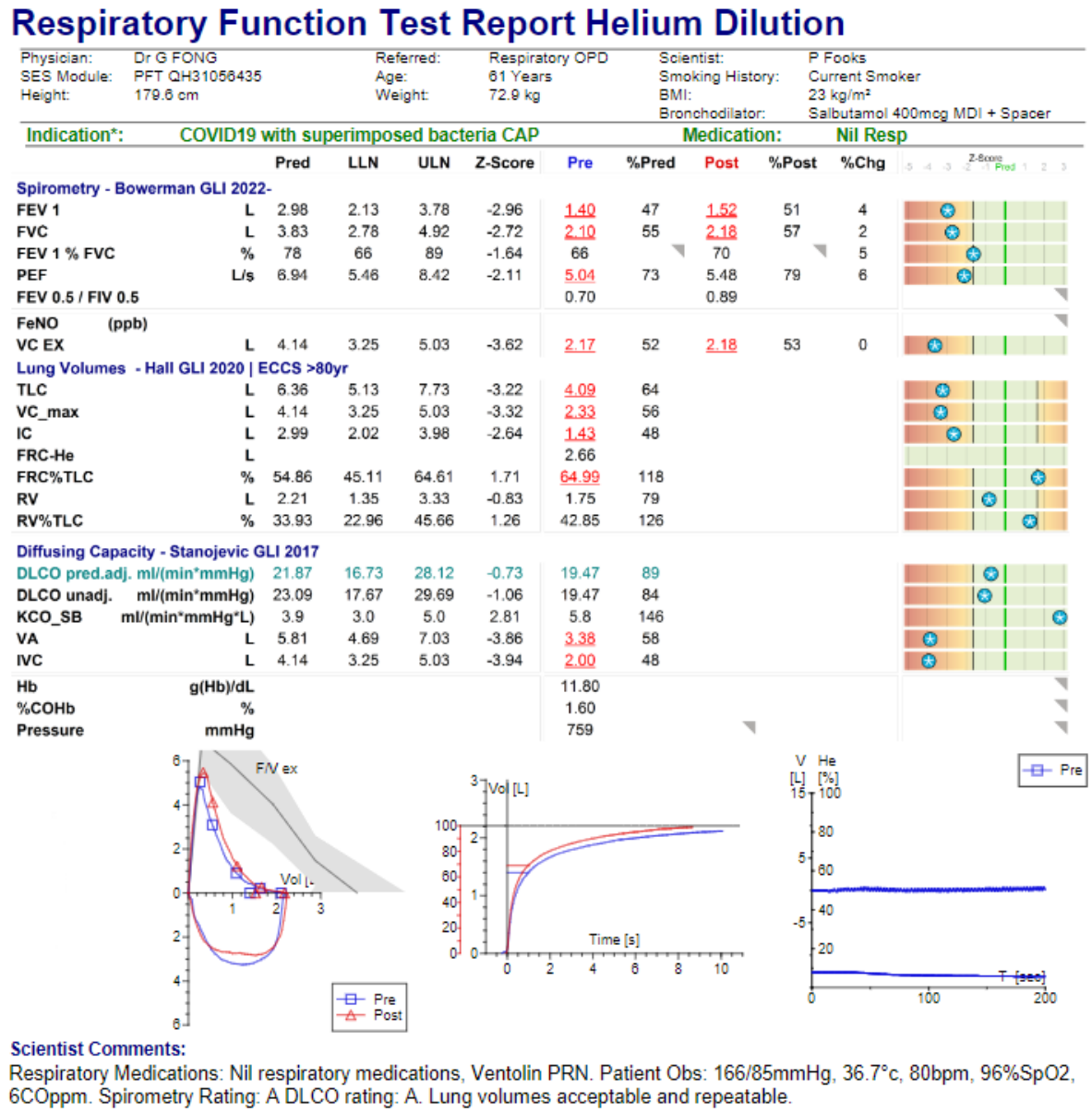
Case Presentation

We present the case of DISH in "Patient M," a 61-year-old Caucasian female who was referred for lung function testing with a productive cough brought on by an extended illness following a cruise in late July. The clinical suspicion was COVID-19 with superimposed bacterial community-acquired pneumonia, and Patient M was hospitalised and treated with nasal O2 therapy, antibiotics and Remdesevir to treat both conditions. As her condition was steadily improving with treatment, she was discharged home after a three-day inpatient stay. The treating respiratory team organised follow-up lung function to assess her post-recovery, and she presented to the respiratory laboratory 6 weeks post-discharge for follow-up testing.

At time of testing, the patient complained to the testing scientist about a chronic cough that was still present, though lessened since her hospital admission. She noted a slow recovery, but otherwise was feeling well. A verbal history was taken by the testing scientist, who noted that Patient M was a current smoker who was prescribed Ventolin by her general practitioner to take on an as-needed basis to help control her cough.

Investigations

A series of lung function tests were requested by Patient M's attending respiratory physician (*Figure 1*), and the patient does not recall any prior lung function. Her testing was performed well, and all acceptability and reversibility criteria are noted to have been met.



Respiratory Function Test Report Helium Dilution

Physician:	Dr G FONG	Referred:	Respiratory OPD	Scientist: P Fooks
SES Module:	PFT QH31056435	Age:	61 Years	Smoking History: Current Smoker
Height:	179.6 cm	Weight:	72.9 kg	BMI: 23 kg/m²
				Bronchodilator: Salbutamol 400mcg MDI + Spacer

Indication*: COVID19 with superimposed bacteria CAP — Medication: Nil Resp

		Pred	LLN	ULN	Z-Score	Pre	%Pred	Post	%Post	%Chg
Spirometry - Bowerman GLI 2022-										
FEV 1	L	2.98	2.13	3.78	-2.96	1.40	47	1.52	51	4
FVC	L	3.83	2.78	4.92	-2.72	2.10	55	2.18	57	2
FEV 1 % FVC	%	78	66	89	-1.64	66		70		5
PEF	L/s	6.94	5.46	8.42	-2.11	5.04	73	5.48	79	6
FEV 0.5 / FIV 0.5						0.70		0.89		
FeNO (ppb)										
VC EX	L	4.14	3.25	5.03	-3.62	2.17	52	2.18	53	0
Lung Volumes - Hall GLI 2020 | ECCS >80yr										
TLC	L	6.36	5.13	7.73	-3.22	4.09	64			
VC_max	L	4.14	3.25	5.03	-3.32	2.33	56			
IC	L	2.99	2.02	3.98	-2.64	1.43	48			
FRC-He	L					2.66				
FRC%TLC	%	54.86	45.11	64.61	1.71	64.99	118			
RV	L	2.21	1.35	3.33	-0.83	1.75	79			
RV%TLC	%	33.93	22.96	45.66	1.26	42.85	126			
Diffusing Capacity - Stanojevic GLI 2017										
DLCO pred.adj. ml/(min*mmHg)		21.87	16.73	28.12	-0.73	19.47	89			
DLCO unadj. ml/(min*mmHg)		23.09	17.67	29.69	-1.06	19.47	84			
KCO_SB ml/(min*mmHg*L)		3.9	3.0	5.0	2.81	5.8	146			
VA	L	5.81	4.69	7.03	-3.86	3.38	58			
IVC	L	4.14	3.25	5.03	-3.94	2.00	48			
Hb	g(Hb)/dL					11.80				
%COHb	%					1.60				
Pressure	mmHg					759				

Scientist Comments:
Respiratory Medications: Nil respiratory medications, Ventolin PRN. Patient Obs: 166/85mmHg, 36.7°c, 80bpm, 96%SpO2, 6COppm. Spirometry Rating: A DLCO rating: A. Lung volumes acceptable and repeatable.

Figure 1 - Lung Function Test Results

The moderately reduced FEV1 and FVC values with no significant bronchodilator response are indicative of a restrictive pattern, which is reinforced by the moderately reduced lung volume parameters. The normal DLCO with an above-normal KCO in this context would indicate extrapulmonary restriction, which was noted by the reporting physician. X-ray and CT scans of her lungs several months before her hospital presentation did not indicate any pulmonary fibrosis or scarring, and her BMI of 23kg/m^3 would not suggest restriction related to body habitus. While it is possible that a degree of respiratory muscle weakness could be an alternate explanation for the

restrictive pattern, taking into consideration the patient's prior history and investigations would not suggest a more probable cause than DISH.

Figure 2 – MRI imaging of Patient M's anterolateral thoracic and lumbar spine

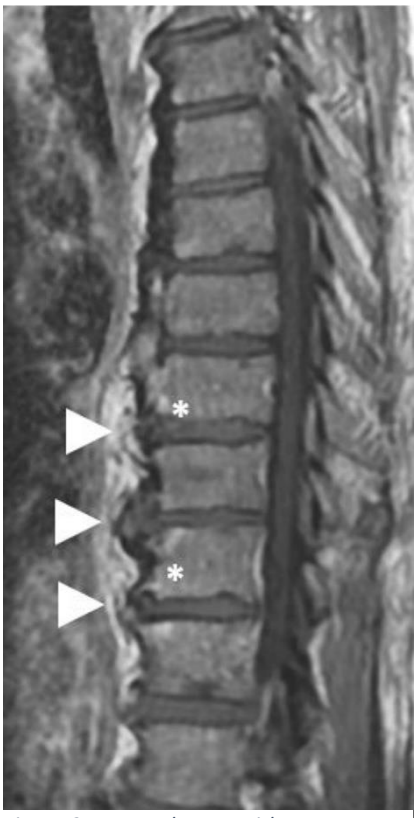


Figure 3 – Example MRI with arrow indication pointing to flowing ossifications characteristic of DISH[11]

When looking at notes from previous clinics that Patient M has attended it becomes apparent that she has been complaining of generalised back pain for a number of years, which increases when she is supine. A previous CT image report from early 2023 noted slight lordosis and kyphosis in the cervical and thoracic spine, respectively. Apparent anterolateral and facet joint osteophytes were noted in the lumbar spine, though an MRI was recommended for further investigation and to differentiate from ankylosing spondylosis, which has a very similar presentation to DISH on X-ray. The MRI found right anterior syndesmophytes "flowing" from the T8 to L1 vertebrae (Figure 2). The presence of ossification visible on both MRI and CT, along with the absence of the typical inflammatory pattern present in ankylosing spondylosis essentially confirms the diagnosis of DISH, and presents a valid explanation for the extrapulmonary restriction seen in the lung function report, due to mechanically limiting the expansion of the chest wall on inspiration[9].

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