patient to the earlier development of diabetes due to factors such as the intrauterine environment of hyperglycaemia.[7] However, none of the factors stated allows us to predict which treatment will be suited to each patient, and this should be based on blood glucose controls and the appearance of complications such as diabetic ketoacidosis.

Nor has it been established what control should be performed in terms of the possibility of the onset of vascular complications. Cases of patients with MODY 6 diabetes and diabetic nephropathy with advanced-stage chronic kidney disease at early ages have been reported, although the greatest severity has been reported in patients with intellectual disabilities, which makes it difficult to establish how their self-care impacted on the development of their condition.[7]

More studies are needed in order to define how diabetes behaves in these patients and the benefits according to the therapies used. The low prevalence and the need for genetic confirmation for its diagnosis make it difficult to gather enough samples to achieve significant results.

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Cystic fibrosis-related diabetes: An interdisciplinary diagnostic and therapeutic challenge

La diabetes relacionada con la fibrosis quística: un reto diagnóstico-terapéutico interdisciplinar

Cystic fibrosis (CF) is the most common autosomal recessive hereditary disease in Caucasian people, with an incidence of one in 3000 live births, and is caused by mutations in the *CFTR* (cystic fibrosis transmembrane conductance regulator) gene.

Abnormal lung function is the main factor responsible for the high mortality rate in patients with CF. However, advances in respiratory therapy and specialised treatment of CF over the last few decades have significantly increased these patients' life expectancy,[1] achieving current mean survival of more than 30 years. This increase in survival has led to an increase in extrapulmonary complications, with cystic fibrosis-related diabetes (CFRD) being the most common comorbidity. CFRD is a comorbidity caused by impairment of the endocrine pancreas and constitutes a determining factor in lung function as well as a marker of a worse prognosis and a higher mortality rate.[2,3] Its prevalence increases with age and its onset is usually preceded even years ahead by carbohydrate metabolism abnormalities, respiratory worsening and weight loss.[4] CFRD may be chronic or intermittent, and it is of multifactorial aetiology. CFRD treatment with insulin appears to improve these patients' respiratory and nutritional status.[5]

Lung transplantation is the only treatment available in end-stage lung disease. The incidence of CFRD in patients who require lung transplantation is 28.6%.[6]

The objectives of the study were to assess the diagnostic possibilities that might be offered by continuous glucose

Table 1 Summary of patients' parameters when starting insulin therapy and changes observed at their six-month follow-up appointment (values specified as ''post'' in the table).

	1	2	3	4	5	6
Gender	♂	♀	♀	♀	♂	♂
Weight percentile	p < 3	p < 3	p37	p26	p31	p29
HbA1c (%)	6.1	6.2	5.9	5.8	5.7	5.7
OGTT	Abnormal	Abnormal	Normal	Abnormal	Abnormal	Abnormal
CGM	Abnormal	Abnormal	Abnormal	Abnormal	Abnormal	Abnormal
Rapid-acting insulin	Yes	Yes	Yes	Yes	Yes	Yes
Insulin glargine	Yes	Yes	No	No	No	No
FVC (%)	67	55	60	55	93	91
FEV1 (%)	56	48	44	43	82	65
FEV1/FVC (%)	70	76	64	66	78	57
Post weight percentile	p3	p9	p61	p37	p46	p41
Post HbA1c (%)	5.6	5.9	5.4	5.4	5.3	5.2
Post FVC (%)	68	100	71.8	91	102	103
Post FEV1 (%)	59	82.4	61.3	69.3	100	100
Post FEV1/FVC (%)	72	82.4	72.6	65	96	97

monitoring (CGM) added to oral glucose tolerance testing (OGTT) in isolation and to analyse respiratory and metabolic improvement brought about by early insulin therapy in these patients.

We report a case series of six patients aged eight to 16 years followed up at our hospital's CF Unit and Diabetes Unit, who received insulin therapy after exhibiting abnormal carbohydrate metabolism by the usual methods (OGTT and glycated haemoglobin [HbA1c]) and CGM, accompanied by nutritional decline and respiratory worsening, in whom improvement following insulin therapy was observed.

All patients had elevated HbA1c levels (≥5.7%); 83.3% of them also had abnormal OGTT (blood glucose >200 mg/dl after two hours). A CGM system was placed and fasting and/or postprandial hyperglycaemia was observed in all cases; postprandial hyperglycaemia alone was seen in the female patient who had had normal OGTT.

Once abnormal carbohydrate metabolism had been confirmed in all patients, insulin therapy was started with multiple doses of subcutaneous rapid-acting insulin in all of them, while two patients were also administered insulin glargine. Personalised diabetes education regarding insulin therapy, self-management, portion control and decompensation was provided. In one case, insulin therapy was stopped after three months due to clinical and spirometric respiratory improvement and suitable weight gain. Currently, the other five cases are still on insulin therapy.

Follow-up was scheduled one month after starting insulin therapy, then after three months, after six months and after a year, in order to monitor changes in nutrition and weight (weight percentile), metabolism (HbA1c) and respiratory function (spirometry). All patients showed respiratory and nutritional improvement in follow-up appointments, with weight gain and an increase in weight percentile (see Table 1). A mean drop in HbA1c levels by 7.3% compared to baseline was seen six months after starting insulin therapy. Follow-up spirometry at six months also showed improvements in the three parameters recorded (forced vital capacity [FVC], forced expiratory volume in one second [FEV1] and FEV1/FVC ratio) in all patients, with mean increases in FVC of 33.38%, in FEV1 of 42.19% and in FEV1/FVC of 19.76%.

Other publications have reported results similar to those seen in our patients, with follow-up showing good metabolic management in CFRD to have documented rapid, prolonged beneficial effects on respiratory decline in CF.[7]

There are case reports of patients with an indication for lung transplantation in whom insulin therapy achieved sufficient improvement to reverse this indication and ward off a need for transplantation for at least another eight years.[7]

The latest guidelines recommend systematic annual OGTT as of 10 years, or earlier in the event of clinical symptoms such as worsening of lung function or nutritional status with no other explanatory cause,[8] in order to detect any abnormality in carbohydrate metabolism as early as possible. In addition, the usefulness of CGM in detecting postprandial hyperglycaemia, which would otherwise go undetected with OGTT, has gained some attention.[8]

Therefore, we believe that CGM could be a useful tool for the early detection of carbohydrate metabolism abnormalities in patients with CF, as well as in cases with normal OGTT but with respiratory and nutritional worsening with no other apparent cause, and patients who require treatment with systemic corticosteroid therapy due to its hyperglycaemia-inducing effects. CGM would therefore enable earlier detection of carbohydrate abnormalities in patients with CF, thereby facilitating earlier initiation of insulin therapy and consequently preventing greater respiratory and metabolic decline.

Conflicts of interest

None.

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Mediastinal paraganglioma: Presurgical embolization

Paraganglioma mediastínico: embolización prequirúrgica

Paragangliomas are neuroendocrine tumours derived from chromaffin cells of the extra-adrenal sympathetic nervous system and they represent an exceptional pathology within the mediastinum, comprising 1–2% of all paragangliomas and less than 0.3% of all mediastinal tumours. The patients' symptoms will depend on functionality and on compressive symptoms because of its large size, although in half of patients it presents as an incidental finding on imaging tests.[1,2]

We present the case study of a functional mediastinal paraganglioma in the middle mediastinum, for which multidisciplinary management was carried out.

The patient was a 55-year-old woman with a personal history of arterial hypertension, diabetes mellitus and dyslipidaemia, who was examined at a private centre for syncope, palpitations and dyspnoea.

The initial study included laboratory tests, chest X-ray, CT scan of the chest and abdomen, and echocardiogram. Following laboratory test findings of catecholamines in 24-h urine: normetanephrine 3,025 μg/24 h (88–444), metanephrine 85 μg/24 h (52–341), elevated chromogranin A (no numerical value available) and imaging (mediastinal large tumour measuring 6 × 7 cm in the middle mediastinum, near the carina, nourished by coronary and bronchial irrigation), mediastinal paraganglioma was diagnosed. At the adrenal glands, no nodules or masses were observed that would suggest a multiple location of the tumour.

The patient was referred to our centre and seen by Endocrinology, who adjusted the alpha-blocker treatment with doxazosin prior to embolisation by Vascular Interventional Radiology (VIR) and subsequent resection by Cardiovascular Surgery (CVS).

A second chest CT scan was performed, which studied the relationship of the lesion to adjacent vascular structures, revealing close contact with the pulmonary veins, left pulmonary artery and left atrium, with irrigation of small arteries from the right coronary artery and left bronchial artery.

The arteriogram performed by VIR showed the tumour irrigation and nutrient vessels that came from multiple arterial branches dependent on the right coronary artery and the left bronchial artery. In the first instance, Onyx 18® (a non-adhesive liquid embolic agent comprising an ethylene vinyl alcohol copolymer dissolved in dimethyl sulfoxide, to which tantalum powder was added for visualisation under fluoroscopy) was used for embolisation of the nutrient branches of the upper pole of the mass from the aorta (bronchial branches), with approach via the right femoral artery. Then, the atrial branches that perfused the lower pole of the tumour (coronary branches) were embolised with Onyx 18® through cardiac catheterisation, with approach via the radial artery. There were no complications during the procedure.

Subsequently (24 h later), surgical excision was performed, planned using a 3D model. A median sternotomy and ligation of multiple arterial and venous tributaries from the descending aorta and coronary arteries were performed, as well as dissection of the area adhered to the left coronary artery and the left atrial roof, with complete resection of the tumour achieved without the need for extracorporeal circulation. The patient had a favourable course after surgery, with no signs of infection or tumour remnants.

Pathology results described an orange specimen with violet-coloured areas and a smooth surface, positive for chromogranin, enolase, synaptophysin, CD56, S100 and vimentin, consistent with a WHO grade I paraganglioma (Fig. 1).

After the procedure, a genetic study of familial paraganglioma with SDHB mutation was conducted, and the result