Images from ^{18}F -DOPA Scan in Congenital Hyperinsulinism: Not Always a Clue for Diagnosis

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Abstract Congenital hyperinsulinism (CHI) is the most common cause of persistent hypoglycaemia in childhood (Horm Res 70:65–72, 2008; J Clin Endocr Metab 93:869–875, 2008). ^{18}F -Fluoro-L-dihydroxy-phenylalanine (^{18}F -DOPA) positron emission tomography (PET) can detect areas of increased activity in the pancreas and may differentiate focal from diffuse CHI (J Clin Endocr Metab 93:869–875, 2008; Radiology 253:216–222, 2009). We here report the case of a girl who complained of recurrent episodes of severe hypoglycaemia despite previous partial pancreatectomy. To evaluate the need for additional surgical intervention, we performed ^{18}F -DOPA PET/computed tomography (CT), which showed a focal lesion corresponding to the anatomical region of the pancreatic

tail. On the other hand, abdominal magnetic resonance imaging (MRI) clearly demonstrated that the ^{18}F -DOPA uptake was in a loop of bowel occupying the previous surgical bed. Our case highlights that bowel uptake can be a possible pitfall in the interpretation of ^{18}F -DOPA PET/CT in children affected by CHI, suggesting that when ^{18}F -DOPA PET/CT results do not fit the clinical picture, magnetic resonance imaging (MRI) may allow a more accurate correlation of the radiotracer activity with the underlying anatomical or pathological structure.

Keywords Congenital hyperinsulinism · Positron emission tomography · Pancreatectomy · Magnetic resonance imaging

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Fig. 1 The child was referred to our clinic when she was 13 months old because of diazoxide-unresponsive congenital hyperinsulinism (CHI) due to a paternally inherited mutation in the *ABCC8* gene. The diagnosis of CHI was made during the first month of life. Due to the persistence of severe hypoglycaemic events despite therapy with diazoxide and octreotide, a partial pancreatectomy was performed when she was 3 months old in her developing country of origin. After surgery, she presented recurrent episodes of severe hypoglycaemia despite maximal medical therapy. To evaluate the need for additional surgical intervention,

we performed ^{18}F -Fluoro-L-dihydroxy-phenylalanine positron emission tomography/computed tomography (^{18}F -DOPA PET/CT). ^{18}F -DOPA PET/CT suggested a focal lesion possibly in the anatomical region of the pancreatic tail, with normal biodistribution of ^{18}F -DOPA in the liver, gallbladder and pelvicalyceal systems of both kidneys [4]. **a** Fused ^{18}F -DOPA PET/CT axial section scan at the level of the pancreatic tail/body; **b** fused coronal section scan; **c** fused sagittal section scan; **d** CT axial section scan at the level of the pancreas; **e** ^{18}F -DOPA PET maximum intensity projection (MIP) image

Fig. 2 T1-weighted abdominal magnetic resonance imaging, performed to elucidate the ^{18}F -DOPA PET/CT findings, showed that only the pancreatic head was preserved with pancreatic parenchyma terminating at the isthmus level. In the region of the pancreatic head, normal signal intensity—similar to that of the liver—is visible in continuity with the pancreatic parenchyma. In the region of the body and tail of the pancreas, there is no evidence of the same signal intensity owing to the absence of pancreatic parenchyma at this level as a consequence of previous surgical resection. In light of these results, additional surgery was not performed. Diazoxide was stopped and oral nifedipine (0.5 mg/kg/day) was added to the treatment, determining a significant improvement of glycaemic control. The patient is now 30 months old and she is in a state of good glycaemic control with no severe hypoglycaemic episodes. She has developed well but has been suffering from intermittent seizures

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Compliance with Ethical Standards

Conflict of Interest Evelina Maines, Luca Giacomello, Mirko D’Onofrio, Matteo Salgarello, Rossella Gaudino, Laura Baggio and Andrea Bordugo declare that they have no conflicts of interests.

Ethical Statement The study has been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments. Written informed consent was obtained from the patient’s mother for publication of this report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal. The identity of the patient has been protected.

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