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INTERNATIONAL COMMUNICATION OFFICE OF PEDIATRIC ENDOCRINE SOCIETIES

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NEWSLETTER

Monogenic diabetes: a proposal of classification with practical implications for treatment

Over the last 15 years, novel insights have been gained in identifying monogenic β -cell diabetes with important implications for clinical management. It accounts for only 1–2% of diabetes cases, but it is often misdiagnosed as either type 1 or type 2. In this type of diabetes, the insulin secretion is reduced or low normal and insulin dependence is not always present. The feeling is that the worldwide prevalence is underestimated and novel insights are forthcoming.

*A recent review published in *Nature Clinical Practice Endocrinology & Metabolism* proposes to replace the old classification of both MODY, based just on aetiology, and neonatal diabetes with a new classification which takes into account genetic aetiology, clinical features and possibility of treatment. This proposal tries to simplify the diagnosis making and establish a treatment flow-chart for most of types of monogenic diabetes. The old label Permanent Neonatal Diabetes Mellitus (PNDM) due to mutations in the Kir6.2 or in the SUR1 subunit of the ATP-sensitive potassium channel can be replaced with a more appropriate K_{ATP} PNDM. In this case, the potassium channel is impaired, with reduced insulin secretion which can be usually restored with high dose of sulfonylurea. Up to date, at least 6 different MODYs are known according to the genetic aetiology. The classification as glucokinase (GCK) and Transcription Factors (TF) DMs would consider aetiology, clinical features and treatment.*

While the first would include all the patients with GCK gene mutations, characterized by mild hyperglycaemia since birth, the second would include all patients with mutations in TF and without extra-pancreatic features, characterized by high levels of blood glucose since teens. In both cases, insulin treatment is not necessary, while a treatment with low dose of sulfonylurea can be beneficial to improve the glycemic control exclusively in TF DM. On the other hand, the presence of extra-pancreatic features, such as deafness (mitochondrial diabetes) or renal disease (MODY5), is suggestive of insulin requirement since the early stage of disease. This classification seems easy to apply and could provide a simple flow-chart for physicians for the choice of the most appropriate treatment. See last page for the practical flow chart.

This is the first newsletter of my tenure as COPES Coordinator. If you have any idea or comment on how to improve the COPES newsletter, please let me know. I would like to thank all those who contributed to COPES and encourage them and others to continue their support in this ongoing effort. Special thanks to Ragnar Hanas for his ongoing collaboration and to Jeff Hitchcock from Children with Diabetes for hosting the website and Novo-Nordisk for sponsoring the printed newsletter.

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We are always in search of items of interest to the international community for inclusion in this newsletter and on the COPES website (www.COPESinternational.org). Contact the Coordinator or Vice Coordinator with your suggestions.

Upcoming Events

Dates and locations in **bold** are annual meetings of the affiliated societies.

2008

August 12-16 Durban, South Africa	34th Annual Meeting of ISPAD Contact: Kuben Pillay West Ville Hospital Westville 7 Spine Road, Suite 561 IDurban 3600 4013 South Africa Tel: +27 31 2655377 Fax: +27 31 2655378 kubenpillay@worldonline.co.za www.ispad2008.com
SEPTEMBER 7-11 Rome, Italy	44th EASD Annual Meeting www.easd2008.com
SEPTEMBER 20-23 Istanbul, Turkey	47th ESPE Meeting Contact: Atilla Buyukgebiz Prof Dokuz Eylul Faculty of Medicine Dept of Paediatric Endocrinology and Adolescence Inciralti, IZMIR, TR-35340, Turkey Tel: +90 232 278 8411 / 90 232 464 4540 Fax: +90 232 278 8411 / 90 232 464 9240 E-mail: atilla.buyukgebiz@gmail.com www.congrex.com/espe2008
OCTOBER 2-4 Yonago, Tottori, Japan	42nd Annual Meeting of JSPE Contact: Susumu Kanzaki, MD, PhD Department of Pediatrics & Perinatology Tottori University Faculty of Medicine 36-1 Yonago-shi Nishimachi Tottori 683-8504, Japan Tel: +81-859-38-6557 Fax: +81-859-38-6559 E-mail: smkanzak@grape.med.tottori-u.ac.jp
OCTOBER 11-14 Lima, Peru	XX SLEP meeting Contact: Dr. Miguel Chavez E-mail: mchavez@upch.edu.pe
OCTOBER 29 - NOVEMBER 1 Seoul, Korea	5th Biennial APPES Scientific Meeting Email: appes@willorganise.com.au
NOVEMBER 5-7 Swansea, Wales	Autumn meeting of the BSPED
NOVEMBER 8-12 Rio de Janeiro, Brazil	13th International Congress of Endocrinology Contact: Ruth Clapauch E-mail: rclapauch@uol.com.br www.ice2008rio.com/
NOVEMBER 17-19 Canberra, Australia	APEG Annual Scientific Meeting E-mail: apegasm@willorganise.com.au www.apeg.org.au

Upcoming Events

Dates and locations in **bold** are annual meetings of the affiliated societies.

2009

SEPTEMBER 9-12 New York, USA	8th Joint ESPE/LWPES Meeting Contact: Paul Saenger Fax: +856.439.0525 E-mail: phsaenger@aol.com and lwpe-espe2009@ahint.com www.lwpe-espe2009.org
SEPTEMBER 16-19 Ljubljana, Slovenia	34th Annual Meeting of ISPAD Contact: Tadej Battelino, MD, PhD E-mail: tadej.battelino@mf.uni-lj.si
SEPTEMBER 26-OCTOBER 1 Vienna, Austria	44th EASD Annual Meeting www.easd.org/customfiles/easd/45th/45th-welcome.html
OCTOBER 1-3 Tochigi, Japan	43rd Annual Meeting of the JSPE Contact: Osamu Arisaka, MD E-mail: arisaka@dokkyomed.ac.jp

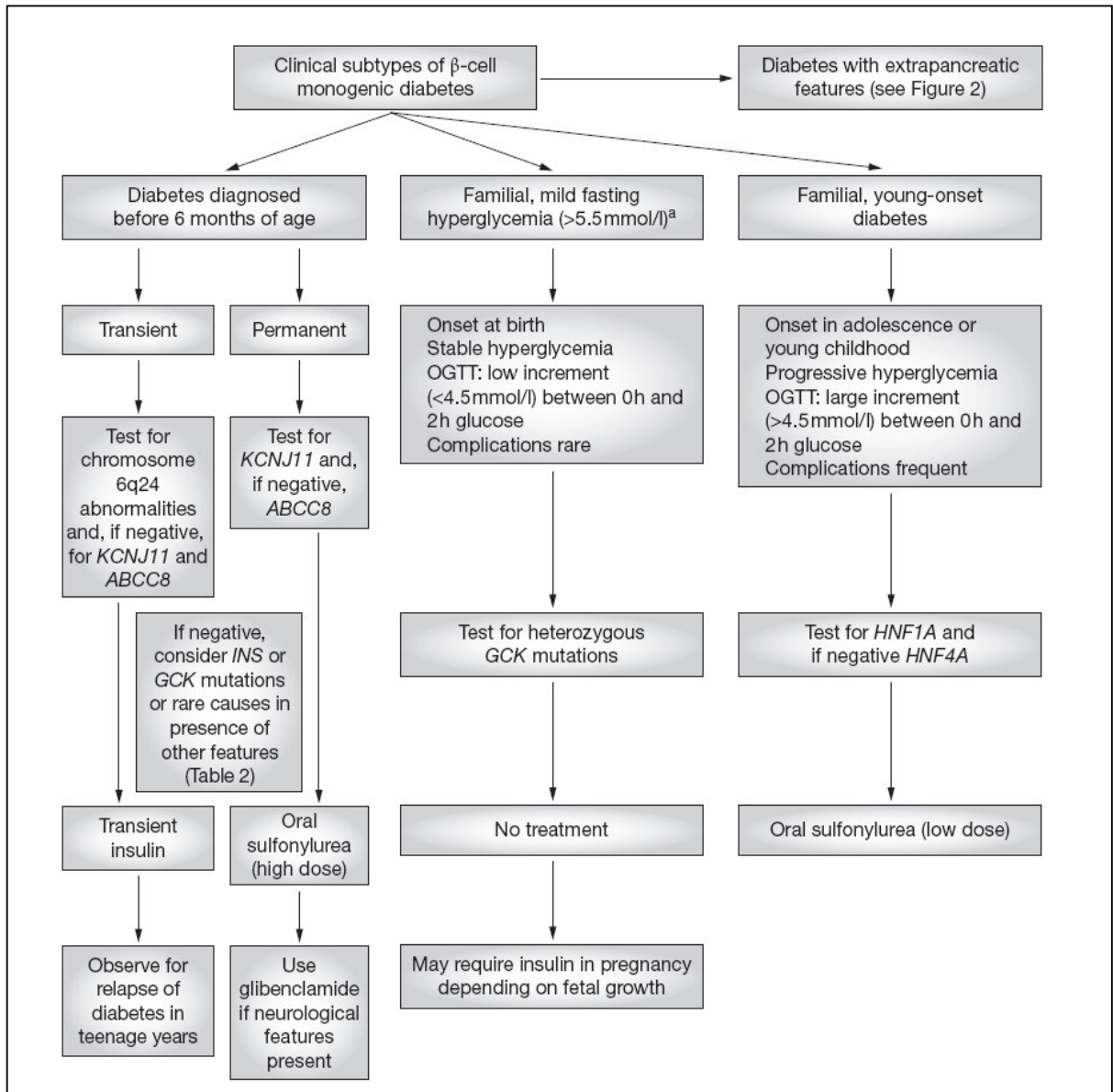
2010

SEPTEMBER 5-11 Buenos Aires, Argentina	35th Annual Meeting of ISPAD Contact: Olgar Ramos, MD E-mail: ramoso@interlink.com.ar
SEPTEMBER 22-25 Prague, Czech Republic	49th ESPE Meeting

2011

SEPTEMBER 28-October 1 Glasgow, Scotland	50th ESPE Meeting
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For more international events, visit www.endo-society.org/apps/Events.



Clinical Implications of a Molecular Genetic Classification of Monogenic Beta-cell Diabetes.
 Nat Clin Pract Endocrinol Metab 2008 Apr;4(4):200-13.

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The COPES newsletter publishes information on the meetings of the affiliated societies and other meetings of interest. Reports on activities of the Affiliated Societies and information on meetings, workshops, courses, fellowships, and exchange programs, as well as selected letters and brief summaries or reviews pertaining to newsworthy items are published in the Newsletter and on the website. All contributions are welcome, so please mail or fax your correspondence to the Coordinator or Vice-Coordinator.

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