

Literaturverzeichnis anderer Autoren

1. Abdel Khalek M, Kandil E. Is octreotide safe for the management of persistent hyperinsulinemic hypoglycemia of infancy? *Eur J Pediatr Surg* 2011;21:188-189
2. Abdulhadi-Atwan M, Bushman J, Tornovsky-Babaey S, et al. Novel de novo mutation in sulfonyleurea receptor 1 presenting as hyperinsulinism in infancy followed by overt diabetes in early adolescence. *Diabetes* 2008;57:1935-1940
3. Adzick NS, Thornton PS, Stanley CA, Kaye RD, Ruchelli E. A multidisciplinary approach to the focal form of congenital hyperinsulinism leads to successful treatment by partial pancreatectomy. *J Pediatr Surg* 2004;39:270-275
4. Al-Nassar S, Sakati N, Al-Ashwal A, Bin-Abbas B. Persistent hyperinsulinaemic hypoglycaemia of infancy in 43 children: long-term clinical and surgical follow-up. *Asian J Surg* 2006;29:207-211
5. Al-Shanafey S. Laparoscopic vs open pancreatectomy for persistent hyperinsulinemic hypoglycemia of infancy. *J Pediatr Surg* 2009;44:957-961
6. Al-Shanafey S, Alkhudhur H. Food aversion among patients with persistent hyperinsulinemic hypoglycemia of infancy. *J Pediatr Surg* 2012;47:895-897
7. Al-Shanafey S, Habib Z, AlNassar S. Laparoscopic pancreatectomy for persistent hyperinsulinemic hypoglycemia of infancy. *J Pediatr Surg* 2009;44:134-138; discussion 138
8. Anlauf M, Wieben D, Perren A, et al. Persistent hyperinsulinemic hypoglycemia in 15 adults with diffuse nesidioblastosis: diagnostic criteria, incidence, and characterization of beta-cell changes. *Am J Surg Pathol* 2005;29:524-533

9. Arbizu Lostao J, Fernandez-Marmiesse A, Garrastachu Zumarran P, et al. [18F-fluoro-L-DOPA PET-CT imaging combined with genetic analysis for optimal classification and treatment in a child with severe congenital hyperinsulinism]. *An Pediatr (Barc)* 2008;68:481-485
10. Arnoux JB, Verkarre V, Saint-Martin C, et al. Congenital hyperinsulinism: current trends in diagnosis and therapy. *Orphanet J Rare Dis* 2011;6:63
11. Avatapalle B, Padidela R, Randell T, Banerjee I. Drug-induced hepatitis following use of octreotide for long-term treatment of congenital hyperinsulinism. *BMJ Case Rep* 2012;2012
12. Aynsley-Green A, Hussain K, Hall J, et al. Practical management of hyperinsulinism in infancy. *Arch Dis Child Fetal Neonatal Ed* 2000;82:F98-F107
13. Bakker B, Oostdijk W. Diagnosis and management of congenital hyperinsulinism: a case report. *Eur J Endocrinol* 2006;155:153-155
14. Balan KK. Visualization of the gall bladder on F-18 FDOPA PET imaging: a potential pitfall. *Clin Nucl Med* 2005;30:23-24
15. Balasubramaniam S, Kapoor R, Yeow JH, et al. Biochemical evaluation of an infant with hypoglycemia resulting from a novel de novo mutation of the GLUD1 gene and hyperinsulinism-hyperammonemia syndrome. *J Pediatr Endocrinol Metab* 2011;24:573-577
16. Banerjee I, Avatapalle B, Padidela R, et al. Integrating genetic and imaging investigations into the clinical management of congenital hyperinsulinism. *Clin Endocrinol (Oxf)* 2013;78:803-813
17. Banerjee I, Avatapalle B, Petkar A, et al. The association of cardiac ventricular hypertrophy with congenital hyperinsulinism. *Eur J Endocrinol* 2012;167:619-624

18. Banerjee I, Skae M, Flanagan SE, et al. The contribution of rapid KATP channel gene mutation analysis to the clinical management of children with congenital hyperinsulinism. *Eur J Endocrinol* 2011;164:733-740
19. Bas VN, Ozkan M, Zenciroglu A, et al. Seizure due to somatostatin analog discontinuation in a case diagnosed as congenital hyperinsulinism novel mutation. *J Pediatr Endocrinol Metab* 2012;25:553-555
20. Bax KN, van der Zee DC. The laparoscopic approach toward hyperinsulinism in children. *Semin Pediatr Surg* 2007;16:245-251
21. Bax NM, van der Zee DC, de Vroede M, Jansen M, Nikkels J. Laparoscopic identification and removal of focal lesions in persistent hyperinsulinemic hypoglycemia of infancy. *Surg Endosc* 2003;17:833
22. Bayarchimeg M, Ismail D, Lam A, et al. Galactokinase deficiency in a patient with congenital hyperinsulinism. *JIMD Rep* 2012;5:7-11
23. Bellanne-Chantelot C, Saint-Martin C, Ribeiro MJ, et al. ABCC8 and KCNJ11 molecular spectrum of 109 patients with diazoxide-unresponsive congenital hyperinsulinism. *J Med Genet* 2010;47:752-759
24. Bellanne-Chantelot C, Saint-Martin C, Ribeiro MJ, et al. ABCC8 and KCNJ11 molecular spectrum of 109 patients with diazoxide-unresponsive congenital hyperinsulinism. *J Med Genet* 2011;47:752-759
25. Beltrand J, Caquard M, Arnoux JB, et al. Glucose metabolism in 105 children and adolescents after pancreatectomy for congenital hyperinsulinism. *Diabetes Care* 2012;35:198-203
26. Ben Turkia H, Brahim K, Azzouz H, et al. Congenital hyperinsulinism: review of 12 Tunisian cases. *Tunis Med* 2011;89:369-373

27. Biagiotti L, Proverbio MC, Bosio L, et al. Identification of two novel frameshift mutations in the KCNJ11 gene in two Italian patients affected by Congenital Hyperinsulinism of Infancy. *Exp Mol Pathol* 2007;83:59-64
28. Blakely ML, Lobe TE, Cohen J, Burghen GA. Laparoscopic pancreatectomy for persistent hyperinsulinemic hypoglycemia of infancy. *Surg Endosc* 2001;15:897-898
29. Bowman P, Flanagan SE, Edghill EL, et al. Heterozygous ABCC8 mutations are a cause of MODY. *Diabetologia* 2011;55:123-127
Branstrom R, Berglund E, Curman P, et al. Electrical short-circuit in beta-cells from a patient with non-insulinoma pancreatogenous hypoglycemic syndrome (NIPHS): a case report. *J Med Case Rep* 2010;4:315
30. Bremer AA, Nobuhara KK, Gitelman SE. Congenital hyperinsulinism in an infant caused by a macroscopic insulin-producing lesion. *J Pediatr Endocrinol Metab* 2007;20:437-440
31. Brunelle F, Negre V, Barth MO, et al. Pancreatic venous samplings in infants and children with primary hyperinsulinism. *Pediatr Radiol* 1989;19:100-103
32. Brunetti-Pierri N, Olutoye OO, Heptulla R, Tatevian N. Case report: pathological features of aberrant pancreatic development in congenital hyperinsulinism due to ABCC8 mutations. *Ann Clin Lab Sci* 2008;38:386-389
33. Bulbul A, Bolat F, Comert S, et al. Persistent hyperinsulinemic hypoglycemia with left ventricular hypertrophy and dysrhythmia: a case report. *Fetal Pediatr Pathol* 2010;29:165-171
34. Burns CM, Rutherford MA, Boardman JP, Cowan FM. Patterns of cerebral injury and neurodevelopmental outcomes after symptomatic neonatal hypoglycemia. *Pediatrics* 2008;122:65-74

35. Capito C, de Lonlay P, Verkarre V, et al. The surgical management of atypical forms of congenital hyperinsulinism. *Semin Pediatr Surg* 2011;20:54-55
36. Capito C, Khen-Dunlop N, Ribeiro MJ, et al. Value of 18F-fluoro-L-dopa PET in the preoperative localization of focal lesions in congenital hyperinsulinism. *Radiology* 2009;253:216-222
37. Casanova D, Polavieja MG, Naranjo A, et al. Surgical treatment of persistent hyperinsulinemic hypoglycemia (PHH) (insulinoma and nesidioblastosis). *Langenbecks Arch Surg* 2007;392:663-670
38. Cherian MP, Abduljabbar MA. Persistent hyperinsulinemic hypoglycemia of infancy (PHHI): Long-term outcome following 95% pancreatectomy. *J Pediatr Endocrinol Metab* 2005;18:1441-1448
39. Cherubini V, Bagalini LS, Ianilli A, et al. Rapid genetic analysis, imaging with 18F-DOPA-PET/CT scan and laparoscopic surgery in congenital hyperinsulinism. *J Pediatr Endocrinol Metab* 2010;23:171-177
40. Chigot V, De Lonlay P, Nassogne MC, et al. Pancreatic arterial calcium stimulation in the diagnosis and localisation of persistent hyperinsulinemic hypoglycaemia of infancy. *Pediatr Radiol* 2001;31:650-655
41. Chik KK, Chan CW, Lam CW, Ng KL. Hyperinsulinism and hyperammonaemia syndrome due to a novel missense mutation in the allosteric domain of the glutamate dehydrogenase 1 gene. *J Paediatr Child Health* 2008;44:517-519
42. Christesen HB, Brusgaard K, Alm J, et al. Rapid genetic analysis in congenital hyperinsulinism. *Horm Res* 2007;67:184-188
43. Christesen HB, Tribble ND, Molven A, et al. Activating glucokinase (GCK) mutations as a cause of medically responsive congenital hyperinsulinism: prevalence in

children and characterisation of a novel GCK mutation. *Eur J Endocrinol* 2008;159:27-34

44. Christesen HT, Brusgaard K, Hussain K. Recurrent spontaneous hypoglycaemia causes loss of neurogenic and neuroglycopenic signs in infants with congenital hyperinsulinism. *Clin Endocrinol (Oxf)* 2011;76:548-554
45. Correa-Giannella ML, Freire DS, Cavaleiro AM, et al. Hyperinsulinism/hyperammonemia (HI/HA) syndrome due to a mutation in the glutamate dehydrogenase gene. *Arq Bras Endocrinol Metabol* 2012;56:485-489
46. Cosgrove KE, Antoine MH, Lee AT, et al. BPDZ 154 activates adenosine 5'-triphosphate-sensitive potassium channels: in vitro studies using rodent insulin-secreting cells and islets isolated from patients with hyperinsulinism. *J Clin Endocrinol Metab* 2002;87:4860-4868
47. Cosgrove KE, Shepherd RM, Fernandez EM, et al. Genetics and pathophysiology of hyperinsulinism in infancy. *Horm Res* 2004;61:270-288
48. Cosgrove KE, Straub SG, Barnes PD, et al. Y-26763: ATP-sensitive K⁺ channel activation and the inhibition of insulin release from human pancreatic beta-cells. *Eur J Pharmacol* 2004;486:133-139
49. Craver RD, Hill CB. Cure of hypoglycemic hyperinsulinism by enucleation of a focal islet cell adenomatous hyperplasia. *J Pediatr Surg* 1997;32:1526-1527
50. Cretolle C, de Lonlay P, Sauvat F, et al. [Congenital hyperinsulinism of infancy: surgical treatment in 60 cases of focal form]. *Arch Pediatr* 2005;12:258-263
51. Cretolle C, Fekete CN, Jan D, et al. Partial elective pancreatectomy is curative in focal form of permanent hyperinsulinemic hypoglycaemia in infancy: A report of 45 cases from 1983 to 2000. *J Pediatr Surg* 2002;37:155-158

52. Dacou-Voutetakis C, Psychou F, Maniati-Christidis M. Persistent hyperinsulinemic hypoglycemia of infancy: long-term results. *J Pediatr Endocrinol Metab* 1998;11 Suppl 1:131-141
53. Damaj L, le Lorch M, Verkarre V, et al. Chromosome 11p15 paternal isodisomy in focal forms of neonatal hyperinsulinism. *J Clin Endocrinol Metab* 2008;93:4941-4947
54. De Leon DD, Li C, Delson MI, et al. Exendin-(9-39) corrects fasting hypoglycemia in SUR-1^{-/-} mice by lowering cAMP in pancreatic beta-cells and inhibiting insulin secretion. *J Biol Chem* 2008;283:25786-25793
55. De Leon DD, Stanley CA. Mechanisms of Disease: advances in diagnosis and treatment of hyperinsulinism in neonates. *Nat Clin Pract Endocrinol Metab* 2007;3:57-68
56. de Lonlay P, Fournet JC, Rahier J, et al. Somatic deletion of the imprinted 11p15 region in sporadic persistent hyperinsulinemic hypoglycemia of infancy is specific of focal adenomatous hyperplasia and endorses partial pancreatectomy. *J Clin Invest* 1997;100:802-807
57. de Lonlay P, Fournet JC, Touati G, et al. Heterogeneity of persistent hyperinsulinaemic hypoglycaemia. A series of 175 cases. *Eur J Pediatr* 2002;161:37-48
58. de Lonlay P, Giurgea I, Sempoux C, et al. Dominantly inherited hyperinsulinaemic hypoglycaemia. *J Inherit Metab Dis* 2005;28:267-276
59. de Lonlay P, Giurgea I, Touati G, Saudubray JM. Neonatal hypoglycaemia: aetiologies. *Semin Neonatol* 2004;9:49-58

60. de Lonlay P, Simon-Carre A, Ribeiro MJ, et al. Congenital hyperinsulinism: pancreatic [18F]fluoro-L-dihydroxyphenylalanine (DOPA) positron emission tomography and immunohistochemistry study of DOPA decarboxylase and insulin secretion. *J Clin Endocrinol Metab* 2006;91:933-940
61. de Lonlay-Debeney P, Poggi-Travert F, Fournet JC, et al. Clinical features of 52 neonates with hyperinsulinism. *N Engl J Med* 1999;340:1169-1175
62. De Vroede M, Bax NM, Brusgaard K, Dunne MJ, Groenendaal F. Laparoscopic diagnosis and cure of hyperinsulinism in two cases of focal adenomatous hyperplasia in infancy. *Pediatrics* 2004;114:e520-522
63. Dekelbab BH, Sperling MA. Recent advances in hyperinsulinemic hypoglycemia of infancy. *Acta Paediatr* 2006;95:1157-1164
64. Delonlay P, Simon A, Galmiche-Rolland L, et al. Neonatal hyperinsulinism: clinicopathologic correlation. *Hum Pathol* 2007;38:387-399
65. Dissanayake AS, Jones V, Fernando DJ. Adult hyperinsulinaemic hypoglycaemia caused by coexisting nesidioblastosis and insulinoma. *Eur J Intern Med* 2008;19:303
66. Dubois J, Brunelle F, Touati G, et al. Hyperinsulinism in children: diagnostic value of pancreatic venous sampling correlated with clinical, pathological and surgical outcome in 25 cases. *Pediatr Radiol* 1995;25:512-516
67. Dunne MJ, Cosgrove KE, Shepherd RM, Aynsley-Green A, Lindley KJ. Hyperinsulinism in infancy: from basic science to clinical disease. *Physiol Rev* 2004;84:239-275

68. Faletra F, Snider K, Shyng SL, et al. Co-inheritance of two ABCC8 mutations causing an unresponsive congenital hyperinsulinism: clinical and functional characterization of two novel ABCC8 mutations. *Gene* 2012;516:122-125
69. Fekete CN, de Lonlay P, Jaubert F, et al. The surgical management of congenital hyperinsulinemic hypoglycemia in infancy. *J Pediatr Surg* 2004;39:267-269
70. Fernandez-Alvarez JR, Rabe H, Wilichowski E, Pekrun A. Pitfalls bei der Diagnose des kongenitalen Hyperinsulinismus: Ein Fallbericht und Übersicht der Literatur. *Klin Pädiatr* 2006;218:233-236
71. Flanagan SE, Kapoor RR, Banerjee I, et al. Dominantly acting ABCC8 mutations in patients with medically unresponsive hyperinsulinaemic hypoglycaemia. *Clin Genet* 2011;79:582-587
72. Flanagan SE, Kapoor RR, Hussain K. Genetics of congenital hyperinsulinemic hypoglycemia. *Semin Pediatr Surg* 2011;20:13-17
73. Flanagan SE, Kapoor RR, Mali G, et al. Diazoxide-responsive hyperinsulinemic hypoglycemia caused by HNF4A gene mutations. *Eur J Endocrinol* 2010;162:987-992
74. Flanagan SE, Xie W, Caswell R, et al. Next-generation sequencing reveals deep intronic cryptic ABCC8 and HADH splicing founder mutations causing hyperinsulinism by pseudoexon activation. *Am J Hum Genet* 2013;92:131-136
75. Fournet JC, Mayaud C, de Lonlay P, et al. Unbalanced expression of 11p15 imprinted genes in focal forms of congenital hyperinsulinism: association with a reduction to homozygosity of a mutation in ABCC8 or KCNJ11. *Am J Pathol* 2001;158:2177-2184

76. Fournier SH, Stanley CA, Kelly A. Protein-sensitive hypoglycemia without leucine sensitivity in hyperinsulinism caused by K(ATP) channel mutations. *J Pediatr* 2006;149:47-52
77. Girard CA, Wunderlich FT, Shimomura K, et al. Expression of an activating mutation in the gene encoding the KATP channel subunit Kir6.2 in mouse pancreatic beta cells recapitulates neonatal diabetes. *J Clin Invest* 2009;119:80-90
78. Giurgea I, Bellanne-Chantelot C, Ribeiro M, et al. Molecular mechanisms of neonatal hyperinsulinism. *Horm Res* 2006;66:289-296
79. Giurgea I, Laborde K, Touati G, et al. Acute insulin responses to calcium and tolbutamide do not differentiate focal from diffuse congenital hyperinsulinism. *J Clin Endocrinol Metab* 2004;89:925-929
80. Giurgea I, Ribeiro MJ, Boddaert N, et al. [Congenital hyperinsulinism in newborn and infant]. *Arch Pediatr* 2005;12:1628-1635
81. Giurgea I, Sanlaville D, Fournet JC, et al. Congenital hyperinsulinism and mosaic abnormalities of the ploidy. *J Med Genet* 2006;43:248-254
82. Giurgea I, Sempoux C, Bellanne-Chantelot C, et al. The Knudson's two-hit model and timing of somatic mutation may account for the phenotypic diversity of focal congenital hyperinsulinism. *J Clin Endocrinol Metab* 2006;91:4118-4123
83. Giurgea I, Ulinski T, Touati G, et al. Factitious hyperinsulinism leading to pancreatectomy: severe forms of Munchausen syndrome by proxy. *Pediatrics* 2005;116:e145-148
84. Glaser B. Lessons in human biology from a monogenic pancreatic beta cell disease. *J Clin Invest* 2011;121:3821-3825

85. Glaser B, Blech I, Krakinovsky Y, et al. ABCC8 mutation allele frequency in the Ashkenazi Jewish population and risk of focal hyperinsulinemic hypoglycemia. *Genet Med* 2011
86. Glaser B, Hirsch HJ, Landau H. Persistent hyperinsulinemic hypoglycemia of infancy: long-term octreotide treatment without pancreatectomy. *J Pediatr* 1993;123:644-650
87. Goh BK, Ooi LL, Cheow PC, et al. Accurate preoperative localization of insulinomas avoids the need for blind resection and reoperation: analysis of a single institution experience with 17 surgically treated tumors over 19 years. *J Gastrointest Surg* 2009;13:1071-1077
88. Gonzalez-Barroso MM, Giurgea I, Bouillaud F, et al. Mutations in UCP2 in congenital hyperinsulinism reveal a role for regulation of insulin secretion. *PLoS One* 2008;3:e3850
89. Goossens A, Gepts W, Saudubray JM, et al. Diffuse and focal nesidioblastosis. A clinicopathological study of 24 patients with persistent neonatal hyperinsulinemic hypoglycemia. *Am J Surg Pathol* 1989;13:766-775
90. Greer RM, Shah J, Jeske YW, et al. Genotype-phenotype associations in patients with severe hyperinsulinism of infancy. *Pediatr Dev Pathol* 2007;10:25-34
91. Gruppuso PA, DeLuca F, O'Shea PA, Schwartz R. Near-total pancreatectomy for hyperinsulinism. Spontaneous remission of resultant diabetes. *Acta Paediatr Scand* 1985;74:311-315
92. Guerrero-Fernandez J, Gonzalez Casado I, Espinoza Colindres L, Gracia Bouthelie R. [Congenital hyperinsulinism. Review of 22 cases]. *An Pediatr (Barc)* 2006;65:22-31

93. Gussinyer M, Clemente M, Cebrian R, et al. Glucose intolerance and diabetes are observed in the long-term follow-up of nonpancreatectomized patients with persistent hyperinsulinemic hypoglycemia of infancy due to mutations in the ABCC8 gene. *Diabetes Care* 2008;31:1257-1259
94. Hardy OT, Hernandez-Pampaloni M, Saffer JR, et al. Accuracy of [18F]fluorodopa positron emission tomography for diagnosing and localizing focal congenital hyperinsulinism. *J Clin Endocrinol Metab* 2007;92:4706-4711
95. Hardy OT, Hernandez-Pampaloni M, Saffer JR, et al. Diagnosis and localization of focal congenital hyperinsulinism by 18F-fluorodopa PET scan. *J Pediatr* 2007;150:140-145
96. Hardy OT, Hohmeier HE, Becker TC, et al. Functional genomics of the beta-cell: short-chain 3-hydroxyacyl-coenzyme A dehydrogenase regulates insulin secretion independent of K⁺ currents. *Mol Endocrinol* 2007;21:765-773
97. Henquin JC, Nenquin M, Sempoux C, et al. In vitro insulin secretion by pancreatic tissue from infants with diazoxide-resistant congenital hyperinsulinism deviates from model predictions. *J Clin Invest* 2011;121:3932-3942
98. Henquin JC, Rahier J. Pancreatic alpha cell mass in European subjects with type 2 diabetes. *Diabetologia* 2011;54:1720-1725
99. Henquin JC, Sempoux C, Marchandise J, et al. Congenital hyperinsulinism caused by hexokinase I expression or glucokinase-activating mutation in a subset of beta-cells. *Diabetes* 2012;62:1689-1696
100. Henwood MJ, Kelly A, Macmullen C, et al. Genotype-phenotype correlations in children with congenital hyperinsulinism due to recessive mutations of the adenosine triphosphate-sensitive potassium channel genes. *J Clin Endocrinol Metab* 2005;90:789-794

101. Hu S, Xu Z, Yan J, et al. The treatment effect of diazoxide on 44 patients with congenital hyperinsulinism. *J Pediatr Endocrinol Metab* 2012;25:1119-1122
102. Huang T, Kelly A, Becker SA, Cohen MS, Stanley CA. Hypertrophic cardiomyopathy in neonates with congenital hyperinsulinism. *Arch Dis Child Fetal Neonatal Ed* 2013;98:F351-354
103. Huopio H, Otonkoski T, Vauhkonen I, et al. A new subtype of autosomal dominant diabetes attributable to a mutation in the gene for sulfonylurea receptor 1. *Lancet* 2003;361:301-307
104. Hussain K. Congenital hyperinsulinism. *Semin Fetal Neonatal Med* 2005;10:369-376
105. Hussain K. Diagnosis and management of hyperinsulinaemic hypoglycaemia of infancy. *Horm Res* 2008;69:2-13
106. Hussain K. Congenital hyperinsulinism and neonatal diabetes mellitus. *Rev Endocr Metab Disord* 2010;11:155-156
107. Hussain K. Investigations for neonatal hypoglycaemia. *Clin Biochem* 2011;44:465-466
108. Hussain K, Bitner-Glindzicz M, Blaydon D, et al. Infantile hyperinsulinism associated with enteropathy, deafness and renal tubulopathy: clinical manifestations of a syndrome caused by a contiguous gene deletion located on chromosome 11p. *J Pediatr Endocrinol Metab* 2004;17:1613-1621
109. Hussain K, Bryan J, Christesen HT, Brusgaard K, Aguilar-Bryan L. Serum glucagon counterregulatory hormonal response to hypoglycemia is blunted in congenital hyperinsulinism. *Diabetes* 2005;54:2946-2951

110. Hussain K, Cosgrove KE. From congenital hyperinsulinism to diabetes mellitus: the role of pancreatic beta-cell KATP channels. *Pediatr Diabetes* 2005;6:103-113
111. Hussain K, Cosgrove KE, Shepherd RM, et al. Hyperinsulinemic hypoglycemia in Beckwith-Wiedemann syndrome due to defects in the function of pancreatic beta-cell adenosine triphosphate-sensitive potassium channels. *J Clin Endocrinol Metab* 2005;90:4376-4382
112. Hussain K, Flanagan SE, Smith VV, et al. An ABCC8 gene mutation and mosaic uniparental isodisomy resulting in atypical diffuse congenital hyperinsulinism. *Diabetes* 2008;57:259-263
113. Hussain K, Seppanen M, Nanto-Salonen K, et al. The diagnosis of ectopic focal hyperinsulinism of infancy with [18F]-dopa positron emission tomography. *J Clin Endocrinol Metab* 2006;91:2839-2842
114. Ismail D, Kapoor RR, Smith V, et al. The Heterogeneity of Focal Forms of Congenital Hyperinsulinism. *J Clin Endocrinol Metab* 2012;97:E0000-E0000
115. Ismail D, Smith VV, de Lonlay P, et al. Familial focal congenital hyperinsulinism. *J Clin Endocrinol Metab* 2011;96:24-28
116. Ismail D, Werther G. Persistent hyperinsulinaemic hypoglycaemia of infancy: 15 years' experience at the Royal Children's Hospital (RCH), Melbourne. *J Pediatr Endocrinol Metab* 2005;18:1103-1109
117. Jack MM, Greer RM, Thomsett MJ, et al. The outcome in Australian children with hyperinsulinism of infancy: early extensive surgery in severe cases lowers risk of diabetes. *Clin Endocrinol (Oxf)* 2003;58:355-364

118. Jack MM, Walker RM, Thomsett MJ, Cotterill AM, Bell JR. Histologic findings in persistent hyperinsulinemic hypoglycemia of infancy: Australian experience. *Pediatr Dev Pathol* 2000;3:532-547
119. James C, Kapoor RR, Ismail D, Hussain K. The genetic basis of congenital hyperinsulinism. *J Med Genet* 2009;46:289-299
120. Johnson D, Shepherd RM, Gill D, et al. Glucose-dependent modulation of insulin secretion and intracellular calcium ions by GKA50, a glucokinase activator. *Diabetes* 2007;56:1694-1702
121. Kaczirek K, Niederle B. Nesidioblastosis: an old term and a new understanding. *World J Surg* 2004;28:1227-1230
122. Kane C, Lindley KJ, Johnson PR, et al. Therapy for persistent hyperinsulinemic hypoglycemia of infancy. Understanding the responsiveness of beta cells to diazoxide and somatostatin. *J Clin Invest* 1997;100:1888-1893
123. Kane C, Shepherd RM, Squires PE, et al. Loss of functional KATP channels in pancreatic beta-cells causes persistent hyperinsulinemic hypoglycemia of infancy. *Nat Med* 1996;2:1344-1347
124. Kapoor RR, Flanagan SE, Arya VB, et al. Clinical and molecular characterisation of 300 patients with congenital hyperinsulinism. *Eur J Endocrinol* 2013;168:557-564
125. Kapoor RR, Flanagan SE, Ellard S, Hussain K. Congenital hyperinsulinism: marked clinical heterogeneity in siblings with identical mutations in the ABCC8 gene. *Clin Endocrinol (Oxf)* 2011;76:312-313
126. Kapoor RR, Flanagan SE, Fulton P, et al. Hyperinsulinism-hyperammonaemia syndrome: novel mutations in the GLUD1 gene and genotype-phenotype correlations. *Eur J Endocrinol* 2009;161:731-735

127. Kapoor RR, Flanagan SE, James C, et al. Hyperinsulinaemic hypoglycaemia. *Arch Dis Child* 2009;94:450-457
128. Kapoor RR, James C, Flanagan SE, et al. 3-Hydroxyacyl-coenzyme A dehydrogenase deficiency and hyperinsulinemic hypoglycemia: characterization of a novel mutation and severe dietary protein sensitivity. *J Clin Endocrinol Metab* 2009;94:2221-2225
129. Kapoor RR, James C, Hussain K. Advances in the diagnosis and management of hyperinsulinemic hypoglycemia. *Nat Clin Pract Endocrinol Metab* 2009;5:101-112
130. Kapoor RR, James C, Hussain K. Hyperinsulinism in developmental syndromes. *Endocr Dev* 2009;14:95-113
131. Kassem SA, Ariel I, Thornton PS, et al. p57(KIP2) expression in normal islet cells and in hyperinsulinism of infancy. *Diabetes* 2001;50:2763-2769
132. Kassem SA, Ariel I, Thornton PS, Scheimberg I, Glaser B. Beta-cell proliferation and apoptosis in the developing normal human pancreas and in hyperinsulinism of infancy. *Diabetes* 2000;49:1325-1333
133. Kauhanen S, Seppanen M, Minn H, et al. Fluorine-18-L-dihydroxyphenylalanine (18F-DOPA) positron emission tomography as a tool to localize an insulinoma or beta-cell hyperplasia in adult patients. *J Clin Endocrinol Metab* 2007;92:1237-1244
134. Kelly A, Tang R, Becker S, Stanley CA. Poor specificity of low growth hormone and cortisol levels during fasting hypoglycemia for the diagnoses of growth hormone deficiency and adrenal insufficiency. *Pediatrics* 2008;122:e522-528
135. Kenney B, Tormey CA, Qin L, et al. Adult nesidioblastosis. Clinicopathologic correlation between pre-operative selective arterial calcium stimulation studies and post-operative pathologic findings. *JOP* 2008;9:504-511

136. Khen-Dunlop N, Capito C, Valayannopoulos V, et al. Predictive value of postoperative glycosuria after partial elective pancreatectomy in focal congenital hyperinsulinism. *Diabetes Care* 2008;31:e71
137. Kloppel G, Anlauf M, Raffel A, Perren A, Knoefel WT. Adult diffuse nesidioblastosis: genetically or environmentally induced? *Hum Pathol* 2008;39:3-8
138. Knudson AG, Jr. Mutation and cancer: statistical study of retinoblastoma. *Proc Natl Acad Sci U S A* 1971;68:820-823
139. Kork F, Blankenstein O, Mohnike W, Höhne C. Sedierung von Kindern zum PET-CT bei kongenitalem Hyperinsulinismus. *Anaesthesist* 2008;57:1087-1090
140. Kubota A, Yonekura T, Usui N, et al. Two cases of persistent hyperinsulinemic hypoglycemia that showed spontaneous regression and maturation of the Langerhans islets. *J Pediatr Surg* 2000;35:1661-1662
141. Kuhnen P, Marquard J, Ernert A, et al. Long-term lanreotide treatment in six patients with congenital hyperinsulinism. *Horm Res Paediatr* 2012;78:106-112
142. Kumaran A, Kapoor RR, Flanagan SE, Ellard S, Hussain K. Congenital hyperinsulinism due to a compound heterozygous ABCC8 mutation with spontaneous resolution at eight weeks. *Horm Res Paediatr* 2010;73:287-292
143. Kushner JA, Simpson L, Wartschow LM, et al. Phosphatase and tensin homolog regulation of islet growth and glucose homeostasis. *J Biol Chem* 2005;280:39388-39393
144. Laje P, Halaby L, Adzick NS, Stanley CA. Necrotizing enterocolitis in neonates receiving octreotide for the management of congenital hyperinsulinism. *Pediatr Diabetes* 2009;11:142-147

145. Langer P, Bartsch DK, Fendrich V, et al. [Minimal-invasive operative treatment of organic hyperinsulinism]. *Dtsch Med Wochenschr* 2005;130:514-518
146. Le Quan Sang KH, Arnoux JB, Mamoune A, et al. Successful treatment of congenital hyperinsulinism with long-acting release octreotide. *Eur J Endocrinol* 2011;166:333-339
147. Leibowitz G, Kaiser N, Cerasi E. Balancing needs and means: the dilemma of the beta-cell in the modern world. *Diabetes Obes Metab* 2009;11 Suppl 4:1-9
148. Leibowitz G, Weintrob N, Pikarsky A, et al. Normal proinsulin processing despite beta-cell dysfunction in persistent hyperinsulinaemic hypoglycaemia of infancy (nesidioblastosis). *Diabetologia* 1996;39:1338-1344
149. Levitt Katz LE, Satin-Smith MS, Collett-Solberg P, et al. Insulin-like growth factor binding protein-1 levels in the diagnosis of hypoglycemia caused by hyperinsulinism. *J Pediatr* 1997;131:193-199
150. Liem NT, Son TN, Hoan NT. Laparoscopic near-total pancreatectomy for persistent hyperinsulinemic hypoglycemia of infancy: report of two cases. *J Laparoendosc Adv Surg Tech A* 2010;20:115-117
151. Liew CG, Moore H, Ruban L, et al. Human embryonic stem cells: possibilities for human cell transplantation. *Ann Med* 2005;37:521-532
152. Liew CG, Shah NN, Briston SJ, et al. PAX4 enhances beta-cell differentiation of human embryonic stem cells. *PLoS One* 2008;3:e1783
153. Limongelli P, Belli A, Cioffi L, et al. Hepatobiliary and pancreatic: nesidioblastosis. *J Gastroenterol Hepatol* 2012;27:1538

154. Lin YW, Bushman JD, Yan FF, et al. Destabilization of ATP-sensitive potassium channel activity by novel KCNJ11 mutations identified in congenital hyperinsulinism. *J Biol Chem* 2008;283:9146-9156
155. Lin YW, MacMullen C, Ganguly A, Stanley CA, Shyng SL. A novel KCNJ11 mutation associated with congenital hyperinsulinism reduces the intrinsic open probability of beta-cell ATP-sensitive potassium channels. *J Biol Chem* 2006;281:3006-3012
156. Lindley KJ, Dunne MJ. Contemporary strategies in the diagnosis and management of neonatal hyperinsulinaemic hypoglycaemia. *Early Hum Dev* 2005;81:61-72
157. Lindley KJ, Spitz L. Surgery of persistent hyperinsulinaemic hypoglycaemia. *Semin Neonatol* 2003;8:259-265
158. Lovisolo SM, Mendonca BB, Pinto EM, et al. Congenital hyperinsulinism in Brazilian neonates: a study of histology, KATP channel genes, and proliferation of beta cells. *Pediatr Dev Pathol* 2010;13:375-384
159. Macmullen CM, Zhou Q, Snider KE, et al. Diazoxide-unresponsive congenital hyperinsulinism in children with dominant mutations of the beta-cell sulfonylurea receptor SUR1. *Diabetes* 2011;60:1797-1804
160. Mannikko R, Flanagan SE, Sim X, et al. Mutations of the same conserved glutamate residue in NBD2 of the sulfonylurea receptor 1 subunit of the KATP channel can result in either hyperinsulinism or neonatal diabetes. *Diabetes* 2011;60:1813-1822
161. Marhfour I, Moulin P, Marchandise J, et al. Impact of Sur1 gene inactivation on the morphology of mouse pancreatic endocrine tissue. *Cell Tissue Res* 2009;335:505-515

162. Martinac I, Bogovic M, Batinica S, et al. [Congenital hyperinsulinism--novel insights into etiology, diagnosis and treatment]. *Lijec Vjesn* 2012;134:286-292
163. Martinez-Ibanez V, Gussinyer M, Toran N, et al. Pancreatectomy extension in persistent hyperinsulinaemic hypoglycaemia: a new strategy. *Eur J Pediatr Surg* 2002;12:262-266
164. Masue M, Nishibori H, Fukuyama S, et al. Diagnostic accuracy of [(1)(8)F]-fluoro-L-dihydroxyphenylalanine positron emission tomography scan for persistent congenital hyperinsulinism in Japan. *Clin Endocrinol (Oxf)* 2011;75:342-346
165. Mazor-Aronovitch K, Gillis D, Lobel D, et al. Long-term neurodevelopmental outcome in conservatively treated congenital hyperinsulinism. *Eur J Endocrinol* 2007;157:491-497
166. Mazor-Aronovitch K, Landau H, Gillis D. Surgical versus non-surgical treatment of congenital hyperinsulinism. *Pediatr Endocrinol Rev* 2009;6:424-430
167. Menni F, de Lonlay P, Sevin C, et al. Neurologic outcomes of 90 neonates and infants with persistent hyperinsulinemic hypoglycemia. *Pediatrics* 2001;107:476-479
168. Mercimek-Mahmutoglu S, Rami B, Feucht M, et al. Long-term follow-up of patients with congenital hyperinsulinism in Austria. *J Pediatr Endocrinol Metab* 2008;21:523-532
169. Modan-Moses D, Koren I, Mazor-Aronovitch K, Pinhas-Hamiel O, Landau H. Treatment of congenital hyperinsulinism with lanreotide acetate (somatuline autogel). *J Clin Endocrinol Metab* 2011;96:2312-2317
170. Negri G, Puglisi A, Gerevini S, Voci C, Zannini P. Thoracoscopic techniques in the management of benign mediastinal dumbbell tumors. *Surg Endosc* 2001;15:897

171. Obatake M, Mochizuki K, Taura Y, et al. Pancreatic head resection preserving the main pancreatic duct for congenital hyperinsulinism of infancy. *Pediatr Surg Int* 2012;28:935-937
172. Ocal G, Flanagan SE, Hacıhamdioglu B, et al. Clinical characteristics of recessive and dominant congenital hyperinsulinism due to mutation(s) in the ABCC8/KCNJ11 genes encoding the ATP-sensitive potassium channel in the pancreatic beta cell. *J Pediatr Endocrinol Metab* 2011;24:1019-1023
173. Okabayashi T, Shima Y, Sumiyoshi T, et al. Diagnosis and management of insulinoma. *World J Gastroenterol* 2013;19:829-837
174. Otonkoski T, Nanto-Salonen K, Seppanen M, et al. Noninvasive diagnosis of focal hyperinsulinism of infancy with [18F]-DOPA positron emission tomography. *Diabetes* 2006;55:13-18
175. Ouyang D, Dhall D, Yu R. Pathologic pancreatic endocrine cell hyperplasia. *World J Gastroenterol* 2011;17:137-143
176. Palladino AA, Bennett MJ, Stanley CA. Hyperinsulinism in infancy and childhood: when an insulin level is not always enough. *Clin Chem* 2008;54:256-263
177. Palladino AA, Stanley CA. A specialized team approach to diagnosis and medical versus surgical treatment of infants with congenital hyperinsulinism. *Semin Pediatr Surg* 2011;20:32-37
178. Park SE, Flanagan SE, Hussain K, et al. Characterization of ABCC8 and KCNJ11 gene mutations and phenotypes in Korean patients with congenital hyperinsulinism. *Eur J Endocrinol* 2011;164:919-926
179. Patterson ME, Mao CS, Yeh MW, et al. Hyperinsulinism presenting in childhood and treatment by conservative pancreatectomy. *Endocr Pract* 2012;18:e52-56

180. Peranteau WH, Bathaii SM, Pawel B, et al. Multiple ectopic lesions of focal islet adenomatosis identified by positron emission tomography scan in an infant with congenital hyperinsulinism. *J Pediatr Surg* 2007;42:188-192
181. Peranteau WH, Ganguly A, Steinmuller L, et al. Prenatal diagnosis and postnatal management of diffuse congenital hyperinsulinism: a case report. *Fetal Diagn Ther* 2006;21:515-518
182. Petrik J, Pell JM, Arany E, et al. Overexpression of insulin-like growth factor-II in transgenic mice is associated with pancreatic islet cell hyperplasia. *Endocrinology* 1999;140:2353-2363
183. Pierro A, Nah SA. Surgical management of congenital hyperinsulinism of infancy. *Semin Pediatr Surg* 2011;20:50-53
184. Pinney SE, MacMullen C, Becker S, et al. Clinical characteristics and biochemical mechanisms of congenital hyperinsulinism associated with dominant KATP channel mutations. *J Clin Invest* 2008;118:2877-2886
185. Placzkowski KA, Vella A, Thompson GB, et al. Secular trends in the presentation and management of functioning insulinoma at the Mayo Clinic, 1987-2007. *J Clin Endocrinol Metab* 2009;94:1069-1073
186. Powell PD, Bellanne-Chantelot C, Flanagan SE, et al. In vitro recovery of ATP-sensitive potassium channels in beta-cells from patients with congenital hyperinsulinism of infancy. *Diabetes* 2011;60:1223-1228
187. Pratt EB, Yan FF, Gay JW, Stanley CA, Shyng SL. Sulfonylurea receptor 1 mutations that cause opposite insulin secretion defects with chemical chaperone exposure. *J Biol Chem* 2009;284:7951-7959

188. Proks P, Ashcroft FM. Modeling K(ATP) channel gating and its regulation. *Prog Biophys Mol Biol* 2009;99:7-19
189. Przybylik-Mazurek E, Pach D, Hubalewska-Dydejczyk A, et al. [Symptoms and early diagnostic possibilities of pancreatic endocrine cells hyperplasia (nesidioblastosis)]. *Przegl Lek* 2012;69:9-14
190. Quan Y, Barszczyk A, Feng ZP, Sun HS. Current understanding of K ATP channels in neonatal diseases: focus on insulin secretion disorders. *Acta Pharmacol Sin* 2011;32:765-780
191. Rahier J, Guiot Y, Sempoux C. Persistent hyperinsulinaemic hypoglycaemia of infancy: a heterogeneous syndrome unrelated to nesidioblastosis. *Arch Dis Child Fetal Neonatal Ed* 2000;82:F108-112
192. Rahier J, Guiot Y, Sempoux C. Morphologic analysis of focal and diffuse forms of congenital hyperinsulinism. *Semin Pediatr Surg* 2011;20:3-12
193. Rahier J, Sempoux C, Fournet JC, et al. Partial or near-total pancreatectomy for persistent neonatal hyperinsulinaemic hypoglycaemia: the pathologist's role. *Histopathology* 1998;32:15-19
194. Ribeiro MJ, Boddaert N, Bellanne-Chantelot C, et al. The added value of [18F]fluoro-L-DOPA PET in the diagnosis of hyperinsulinism of infancy: a retrospective study involving 49 children. *Eur J Nucl Med Mol Imaging* 2007;34:2120-2128
195. Ribeiro MJ, Boddaert N, Delzescaux T, et al. Functional imaging of the pancreas: the role of [18F]fluoro-L-DOPA PET in the diagnosis of hyperinsulinism of infancy. *Endocr Dev* 2007;12:55-66

196. Ribeiro MJ, De Lonlay P, Delzescaux T, et al. Characterization of hyperinsulinism in infancy assessed with PET and 18F-fluoro-L-DOPA. *J Nucl Med* 2005;46:560-566
197. Rother KI, Matsumoto JM, Rasmussen NH, Schwenk WF. Subtotal pancreatectomy for hypoglycemia due to congenital hyperinsulinism: long-term follow-up of neurodevelopmental and pancreatic function. *Pediatr Diabetes* 2001;2:115-122
198. Ruan W, Lai M. Insulin-like growth factor binding protein: a possible marker for the metabolic syndrome? *Acta Diabetol*;47:5-14
199. Saint-Martin C, Arnoux JB, de Lonlay P, Bellanne-Chantelot C. KATP channel mutations in congenital hyperinsulinism. *Semin Pediatr Surg* 2011;20:18-22
200. Sandal T, Laborie LB, Brusgaard K, et al. The spectrum of ABCC8 mutations in Norwegian patients with congenital hyperinsulinism of infancy. *Clin Genet* 2009;75:440-448
201. Scott-Coombes D, Eatock F, Henley R, Luzio SD. Use of intraoperative insulin assay for the localisation of insulinoma. *World J Surg* 2009;33:969-971
202. Sempoux C, Capito C, Bellanne-Chantelot C, et al. Morphological Mosaicism of the Pancreatic Islets: A Novel Anatomopathological Form of Persistent Hyperinsulinemic Hypoglycemia of Infancy. *J Clin Endocrinol Metab* 2011
203. Sempoux C, Guiot Y, Dahan K, et al. The focal form of persistent hyperinsulinemic hypoglycemia of infancy: morphological and molecular studies show structural and functional differences with insulinoma. *Diabetes* 2003;52:784-794
204. Sempoux C, Guiot Y, Lefevre A, et al. Neonatal hyperinsulinemic hypoglycemia: heterogeneity of the syndrome and keys for differential diagnosis. *J Clin Endocrinol Metab* 1998;83:1455-1461

205. Sempoux C, Guiot Y, Rahier J. The focal form of persistent hyperinsulinemic hypoglycemia of infancy. *Diabetes* 2001;50 Suppl 1:S182-183
206. Service FJ, Nathan DM, Mulder JE. Nonislet cell tumor hypoglycemia. In: UpToDate ed; 2011
207. Service FJ, Natt N, Thompson GB, et al. Noninsulinoma pancreatogenous hypoglycemia: a novel syndrome of hyperinsulinemic hypoglycemia in adults independent of mutations in Kir6.2 and SUR1 genes. *J Clin Endocrinol Metab* 1999;84:1582-1589
208. Service GJ, Thompson GB, Service FJ, et al. Hyperinsulinemic hypoglycemia with nesidioblastosis after gastric-bypass surgery. *N Engl J Med* 2005;353:249-254
209. Shemer R, Avnon Ziv C, Laiba E, et al. Relative expression of a dominant mutated ABCC8 allele determines the clinical manifestation of congenital hyperinsulinism. *Diabetes* 2012;61:258-263
210. Shield JP. Fluorine-18 L-3,4-dihydroxyphenylalanine positron emission tomography: improving surgery and outcome in focal hyperinsulinism. Commentary to Mohnike et al.: Proposal for a standardized protocol for F-DOPA-PET (PET/CT) in congenital hyperinsulinism (*Horm Res* 2006;66:40-42). *Horm Res* 2006;66:43-44
211. Smith VV, Malone M, Risdon RA. Focal or diffuse lesions in persistent hyperinsulinemic hypoglycemia of infancy: concerns about interpretation of intraoperative frozen sections. *Pediatr Dev Pathol* 2001;4:138-143
212. Sperling MA. PET scanning for infants with HHI: a small step for affected infants, a giant leap for the field. *J Pediatr* 2007;150:122-124

213. Spitz L, Bhargava RK, Grant DB, Leonard JV. Surgical treatment of hyperinsulinaemic hypoglycaemia in infancy and childhood. *Arch Dis Child* 1992;67:201-205
214. Stanescu DE, Hughes N, Kaplan B, Stanley CA, De Leon DD. Novel presentations of congenital hyperinsulinism due to mutations in the MODY genes: HNF1A and HNF4A. *J Clin Endocrinol Metab* 2012;97:E2026-2030
215. Stanley CA. Advances in diagnosis and treatment of hyperinsulinism in infants and children. *J Clin Endocrinol Metab* 2002;87:4857-4859
216. Stanley CA. Hypoglycemia in the neonate. *Pediatr Endocrinol Rev* 2006;4 Suppl 1:76-81
217. Stanley CA, Thornton PS, Ganguly A, et al. Preoperative evaluation of infants with focal or diffuse congenital hyperinsulinism by intravenous acute insulin response tests and selective pancreatic arterial calcium stimulation. *J Clin Endocrinol Metab* 2004;89:288-296
218. Stevens A, Cosgrove KE, Padidela R, et al. Can Network biology unravel the aetiology of congenital hyperinsulinism? *Orphanet J Rare Dis* 2013;8
219. Straub SG, Cosgrove KE, Ammala C, et al. Hyperinsulinism of infancy: the regulated release of insulin by KATP channel-independent pathways. *Diabetes* 2001;50:329-339
220. Suchi M, MacMullen C, Thornton PS, et al. Histopathology of congenital hyperinsulinism: retrospective study with genotype correlations. *Pediatr Dev Pathol* 2003;6:322-333

221. Suchi M, MacMullen CM, Thornton PS, et al. Molecular and immunohistochemical analyses of the focal form of congenital hyperinsulinism. *Mod Pathol* 2006;19:122-129
222. Suchi M, Thornton PS, Adzick NS, et al. Congenital hyperinsulinism: intraoperative biopsy interpretation can direct the extent of pancreatectomy. *Am J Surg Pathol* 2004;28:1326-1335
223. Szollosi A, Nenquin M, Aguilar-Bryan L, Bryan J, Henquin JC. Glucose stimulates Ca²⁺ influx and insulin secretion in 2-week-old beta-cells lacking ATP-sensitive K⁺ channels. *J Biol Chem* 2007;282:1747-1756
224. Takaori K, Tanigawa N. Laparoscopic pancreatic resection: the past, present, and future. *Surg Today* 2007;37:535-545
225. Taneja TK, Mankouri J, Karnik R, et al. Sar1-GTPase-dependent ER exit of KATP channels revealed by a mutation causing congenital hyperinsulinism. *Hum Mol Genet* 2009;18:2400-2413
226. Tessonnier L, Sebag F, Ghander C, et al. Limited value of 18F-F-DOPA PET to localize pancreatic insulin-secreting tumors in adults with hyperinsulinemic hypoglycemia. *J Clin Endocrinol Metab* 2009;95:303-307
227. Thakur S, Flanagan SE, Ellard S, Verma IC. Congenital hyperinsulinism caused by mutations in ABCC8 (SUR1) gene. *Indian Pediatr* 2011;48:733-734
228. Thomas PM, Cote GJ, Wohlk N, et al. Mutations in the sulfonylurea receptor gene in familial persistent hyperinsulinemic hypoglycemia of infancy. *Science* 1995;268:426-429

229. Thomas PM, Wohllk N, Huang E, et al. Inactivation of the first nucleotide-binding fold of the sulfonylurea receptor, and familial persistent hyperinsulinemic hypoglycemia of infancy. *Am J Hum Genet* 1996;59:510-518
230. Thompson GB, Service FJ, Andrews JC, et al. Noninsulinoma pancreatogenous hypoglycemia syndrome: an update in 10 surgically treated patients. *Surgery* 2000;128:937-944;discussion 944-935
231. Tornovsky S, Crane A, Cosgrove KE, et al. Hyperinsulinism of infancy: novel ABCC8 and KCNJ11 mutations and evidence for additional locus heterogeneity. *J Clin Endocrinol Metab* 2004;89:6224-6234
232. Vanderveen KA, Grant CS, Thompson GB, et al. Outcomes and quality of life after partial pancreatectomy for noninsulinoma pancreatogenous hypoglycemia from diffuse islet cell disease. *Surgery* 2010;148:1237-1245; discussion 1245-1236
233. Vercellino GF, Cremonese M, Carlando G, et al. Transient neonatal hyperinsulinemic hypoglycemia and neurological outcome: a case report. *Minerva Pediatr* 2011;63:111-114
234. Verheul JC, Ris-Stalpers C, Bikker H, Bakker-van Waarde WM, Noordam C. [Congenital hyperinsulinism in the north-east Netherlands. Clinical features and DNA diagnostics in 22 children]. *Ned Tijdschr Geneesk* 2011;155:A3343
235. Verkarre V, Fournet JC, de Lonlay P, et al. Paternal mutation of the sulfonylurea receptor (SUR1) gene and maternal loss of 11p15 imprinted genes lead to persistent hyperinsulinism in focal adenomatous hyperplasia. *J Clin Invest* 1998;102:1286-1291
236. Warden MJ, German JC, Buckingham BA. The surgical management of hyperinsulinism in infancy due to nesidioblastosis. *J Pediatr Surg* 1988;23:462-465

237. Washington RL, Wolfsdorf JI. Understanding protein-sensitive hypoglycemia. *J Pediatr* 2006;149:6-7
238. Yadav D, Dhingra B, Kumar S, Kumar V, Dutta AK. Persistent hyperinsulinemic hypoglycemia of infancy. *J Pediatr Endocrinol Metab* 2012;25:591-593
239. Yan FF, Casey J, Shyng SL. Sulfonylureas correct trafficking defects of disease-causing ATP-sensitive potassium channels by binding to the channel complex. *J Biol Chem* 2006;281:33403-33413
240. Yang J, Hao R, Zhu X. Diagnostic role of 18F-dihydroxyphenylalanine positron emission tomography in patients with congenital hyperinsulinism: a meta-analysis. *Nucl Med Commun* 2013;34:347-353
241. Yorifuji T, Hosokawa Y, Fujimaru R, et al. Lasting 18F-DOPA PET uptake after clinical remission of the focal form of congenital hyperinsulinism. *Horm Res Paediatr* 2011;76:286-290
242. Yorifuji T, Kawakita R, Hosokawa Y, et al. Efficacy and safety of long-term, continuous subcutaneous octreotide infusion for patients with different subtypes of KATP-channel hyperinsulinism. *Clin Endocrinol (Oxf)* 2012;78:891-897
243. Yorifuji T, Kawakita R, Nagai S, et al. Molecular and clinical analysis of Japanese patients with persistent congenital hyperinsulinism: predominance of paternally inherited monoallelic mutations in the KATP channel genes. *J Clin Endocrinol Metab* 2011;96:E141-145
244. Zhang N, Yang J, Yuan L, et al. 18F-DOPA positron emission tomography/computed tomography application in congenital hyperinsulinism. *J Pediatr Endocrinol Metab* 2012;25:619-622

245. Zhou GW, Wei Y, Chen X, et al. Diagnosis and surgical treatment of multiple endocrine neoplasia. *Chin Med J (Engl)* 2009;122:1495-1500
246. Ziegler AG, Nepom GT. Prediction and pathogenesis in type 1 diabetes. *Immunity* 2010;32:468-478
247. Zschocke J. Dominant versus recessive: molecular mechanisms in metabolic disease. *J Inherit Metab Dis* 2008;31:599-618