contribution—are now widespread, driven by the ever increasing antigenicity of a β -cell upregulated to accommodate ever increasing demands on its function.

The mounting epidemic of obesity has done more than merely test the resources of nations to contain it. By disturbing the control systems that regulate blood glucose—by conducting what amounts in endocrine terms to a population-wide stress test—the Great Weight Gain Experiment has exposed responses that are not consistent with the conventional model of type 1 diabetes. It is not, of course, a perfect experiment, for it lacks the control arm necessary to exclude confounders. But if the changing behaviour of autoantibodies is to be accommodated within a meaningful taxonomy, and the proliferation of “neo-diseases” stemmed, we should maybe think of switching the type 1 diabetes paradigm from immune dysregulation to one of insulin resistance.

Arch Dis Child 2006;**91**:456–458.
doi: 10.1136/adc.2006.094037

Correspondence to: Prof. T J Wilkin, University Medicine, Level 7, Derriford Hospital, Plymouth PL6 8DH, UK; T.Wilkin@pms.ac.uk

Competing interests: none declared

REFERENCES

- 1 Prentice AM. Obesity—the inevitable penalty of civilisation? *Br Med Bull* 1997;**53**:229–37.
- 2 Bundred P, Kitchiner D, Buchan I. Prevalence of overweight and obese children between 1989 and 1998: population based series of cross sectional studies. *BMJ* 2001;**322**:326–8.
- 3 Wilkin TJ, Voss LD. Metabolic syndrome: maladaptation to a modern world. *J R Soc Med* 2004;**97**:511–20.

- 4 Weir GC, Bonner-Weir S. Five stages of evolving beta-cell dysfunction during progression to diabetes. *Diabetes* 2004;**53**(suppl 3):S16–21.
- 5 Silverstein JH, Rosenbloom AL. Type 2 diabetes in children. *Curr Diab Rep* 2001;**1**:19–27.
- 6 Gale EA. The rise of childhood type 1 diabetes in the 20th century. *Diabetes* 2002;**51**:3353–61.
- 7 Kurtz Z, Peckham CS, Ades AE. Changing prevalence of juvenile-onset diabetes mellitus. *Lancet* 1988;**2**:88–90.
- 8 Weets I, De Leeuw IH, Du Caju MV, et al. The incidence of type 1 diabetes in the age group 0–39 years has not increased in Antwerp (Belgium) between 1989 and 2000: evidence for earlier disease manifestation. *Diabetes Care* 2002;**25**:840–6.
- 9 Gilliam LK, Brooks-Worrell BM, Palmer JP, et al. Autoimmunity and clinical course in children with type 1, type 2, and type 1.5 diabetes. *J Autoimmun* 2005;**25**:244–50.
- 10 Molbak AG, Christau B, Marner B, et al. Incidence of insulin-dependent diabetes mellitus in age groups over 30 years in Denmark. *Diabet Med* 1994;**11**:650–5.
- 11 Freedman ZR, Feek CM, Irvine WJ, et al. Islet cell cytoplasmic and cell surface antibodies in diabetes mellitus. *Trans Assoc Am Physicians* 1979;**92**:64–76.
- 12 Irvine WJ, McCallum CJ, Gray RS, et al. Clinical and pathogenic significance of pancreatic-islet-cell antibodies in diabetics treated with oral hypoglycaemic agents. *Lancet* 1977;**1**:1025–7.
- 13 Juneja R, Palmer JP. Type 1½ diabetes: myth or reality? *Autoimmunity* 1999;**29**:65–83.
- 14 Libman IM, Becker DJ. Coexistence of type 1 and type 2 diabetes mellitus: “double” diabetes? *Pediatr Diabetes* 2003;**4**:110–13.
- 15 Reinehr T, Schober E, Wiegand S, et al. β -cell autoantibodies in children with type 2 diabetes mellitus: subgroup or misclassification? *Arch Dis Child* 2006;**91**:473–7.
- 16 American Diabetes Association. Type 2 diabetes in children and adolescents. *Diabetes Care* 2000;**23**:381–9.
- 17 Brooks-Worrell BM, Greenbaum CJ, Palmer JP, et al. Autoimmunity to islet proteins in children diagnosed with new-onset diabetes. *J Clin Endocrinol Metab* 2004;**89**:2222–7.
- 18 Wilkin TJ. The accelerator hypothesis: weight gain as the missing link between type I and type II diabetes. *Diabetologia* 2001;**44**:914–22.
- 19 Onkamo P, Vaananen S, Karvonen M, et al. Worldwide increase of type 1 diabetes—analysis of the data on published incidence trends. *Diabetologia* 1999;**42**:1395–403.
- 20 Marcovecchio M, Mohn A, Chiarelli F. Type 2 diabetes mellitus in children and adolescents. *J Endocrinol Invest* 2005;**28**:853–63.
- 21 Voss LD, Kirkby J, Metcalf BS, et al. Preventable factors in childhood that lead to insulin resistance, diabetes mellitus and the metabolic syndrome: the EarlyBird diabetes study 1. *J Pediatr Endocrinol Metab* 2003;**16**:1211–24.
- 22 Hermann R, Knip M, Veijola R, et al. Temporal changes in the frequencies of HLA genotypes in patients with type 1 diabetes—indication of an increased environmental pressure? *Diabetologia* 2003;**46**:420–5.
- 23 Gillespie KM, Bain SC, Barnett AH, et al. The rising incidence of childhood type 1 diabetes and reduced contribution of high-risk HLA haplotypes.
- 24 Baum JD, Onsted M, Smith MA. Weight gain in infancy and subsequent development of diabetes mellitus in childhood. *Lancet* 1975;**2**:866.
- 25 Johansson C, Samuelsson U, Ludvigsson J. A high weight gain in early life is associated with an increased risk of type 1 (insulin-dependent) diabetes. *Diabetologia* 1994;**37**:91–4.
- 26 Hypponen E, Kenward MG, Virtanen SM, et al. Infant feeding, early weight gain and risk of type 1 diabetes. *Diabetes Care* 1999;**22**:1961–5.
- 27 Kibirige M, Metcalf B, Renuka R, et al. Testing the accelerator hypothesis: the relationship between body mass and age at diagnosis of type 1 diabetes. *Diabetes Care* 2003;**26**:2865–70.
- 28 Betts P, Mulligan J, Ward P, et al. Increasing body weight predicts the earlier onset of insulin-dependant diabetes in childhood: testing the “accelerator hypothesis” (2). *Diabet Med* 2005;**22**:144–51.
- 29 Knerr I, Wolf J, Reinehr T, et al. DPV Scientific Initiative of Germany and Austria: the “accelerator hypothesis”: relationship between weight, height, body mass index and age at diagnosis in a large cohort of 9,248 German and Austrian children with type 1 diabetes mellitus. *Diabetologia* 2005;**48**:2501–4.
- 30 Kordonouri O, Hartmann R. Higher body weight is associated with earlier onset of type 1 diabetes: confirming the accelerator hypothesis. *Diabetic Med* 2005;**22**:1783–4.
- 31 Dabelea D, D’Agostino RB Jr, Mayer-Davis EJ, et al. Testing the accelerator hypothesis: body size, beta-cell function, and age at onset of type 1 (autoimmune) diabetes. *Diabetes Care* 2006;**29**:290–4.
- 32 Fourlanos S, Narendran P, Byrnes GB, et al. Insulin resistance is a risk factor for progression to type 1 diabetes. *Diabetologia* 2004;**47**:1661–7.

Community child health

Balancing biomedical, care, and support needs in the technology dependent child

D W Vickers, L C Maynard

Perspective on the paper by Sullivan *et al* (see page 478)

In this issue, Sullivan *et al* report on their findings of a study of gastrostomy feeding in children with cerebral palsy.¹ This study is part of a larger examination of the impact of this intervention on children. Their key findings are that there is no increase in respiratory

morbidity following gastrostomy insertion and indeed a reduced need for antibiotics or hospital admission in the 12 months following the procedure for children with cerebral palsy. As they point out, there are a number of reports in the literature that have suggested the

reverse finding, an increase in mortality following gastrostomy. A recent systematic review highlighted the continued paucity of evidence on the risks and benefits of this intervention, in particular noting an excess of deaths in the gastrostomy fed group with cerebral palsy, observing however that this may reflect differing levels of disability in the intervention and control groups.²

Clearly there are risks in any surgical procedure, but this paper goes some way to provide reassurance that the medical risks are small. This paper is therefore useful in providing further information for professionals working with children and families to be clearer about the biomedical risks and benefits of this procedure.

This study reports on experience in specialist multidisciplinary feeding clinics for children with neurological disabilities which is likely to mean that they are

reporting on best practice. In general, concentration of expertise and resources in specialist centres results in improved outcomes, but we need to be cautious about assuming that the results are applicable outside of such specialist units.

In our experience it is the often lengthy period of time before arriving at a decision to have a gastrostomy that is problematic. A common concern of families, as the concept of a feeding gastrostomy is initially introduced, is that a gastrostomy will actually increase their burden of care and complicate school placement. However, a number of studies have indicated that this is an exaggerated fear. Sullivan *et al* reported on their experiences in the same clinics and showed improved quality of life after gastrostomy.³ A study of 29 responses in a follow up questionnaire revealed only one parent who regretted the operation.⁴ However other work, particularly those using qualitative methodologies, with families of technology dependent children, including gastrostomies, indicated that there is a significant impact on families. Families reported that technology was demanding of time and impacted on the ability of the family to participate in school, work, and social life.⁵ Families also highlighted the paucity of community based support available, with particular problems around equipment provision, poor planning, and a lack of coordination.⁶ Parents took on complex technical tasks that in hospital would be carried out by trained nurses. This impacted on their parenting and they benefit from support both in technical issues but also with opportunities to discuss their feelings about caring and the impact on their role as parents.⁷ They report that family life is not normal, but can still see the positive and normal aspects of life, while recognising the impact on their family and feel that they can still have a good life.⁸

An important consequence of gastrostomy feeding on children and parents is the loss or reduction of oral feeding. In our experience the positive impact of improved nutrition is well recognised by parents, as is the ease of administering medication. Parents invest a lot of time in feeding their disabled children, and proceeding to a gastrostomy can be seen by them as a failure on their part. We should also not ignore the emotional impact on parents, with the loss of the nurturing experience of feeding. Sleight interviewed parents of children who were both orally and gastrostomy fed; she highlighted the importance placed on oral feeding by mothers, and suggested that support for it should be given a higher priority.⁹ She also cautioned against very early gastrostomy placement unless essential, given the importance of oral feeding in bonding.

Some of the parents in her study commented on the impact of gastrostomy feeding on their interaction with their child; seeing feeding moving from a nurturing to a clinical experience.

Most children with cerebral palsy are cared for by community or neurodisability paediatricians, and referred to surgeons when the decision has been reached to offer gastrostomy. How do parents and professionals arrive at a decision to have a gastrostomy? A study involving 50 mothers in semi-structured interviews on decision making about gastrostomy insertion is revealing.¹⁰ A lack of the right type or amount of information about the impact of the decision they were being asked to make was a common concern. Other concerns were the appearance of the tube, its location on the body, how to insert a tube and to administer feeds, as well as benefits and disadvantages of insertion. A significant number of parents felt unsupported during the decision making process and half reported feeling pressurised by other family members and healthcare professionals. Given that the decision is usually one that takes place over time, rather than as an emergency, in our view we have a duty, both legally and ethically, to ensure that there is sharing of all necessary information, and that parents are enabled to come to a decision in their own timescale and without undue influence from professionals. The evidence from this most recent study is a further help in decision making, as it provides additional reassurance that the medical risk of the procedure is acceptable.

Ethically, the main issue is the balance of beneficence and non-maleficence.¹¹ Does this procedure provide sufficient benefit to outweigh the concerns about morbidity? Sullivan *et al*'s paper suggests that for this particular client group, respiratory morbidity is low.¹ However, it does not address other morbidity, such as impact on family life. In our experience the use of technology to support children at home is increasingly seen by health professionals as normal. We need to consider the impact of these treatments on family functioning. Qualitative studies which have used in-depth interviews with families reveal a mixed picture with both recognition of health gain and real negative impact on family functioning.^{5 7 9}

It is not clear whether the evidence represented here will result in an increased use of this procedure. Gastrostomy feeding seems an appropriate intervention for children with a wide variety of neurological conditions where there are problems with nutrition. However, it needs to be seen in the wider context of impact on family life. This study provides further evidence of improvement in health. However, in our

view, the decision to proceed to a gastrostomy needs to continue to be seen as an important one, with time and information to help parents decide. There needs to be a balance between the biomedical needs of the child and the social impact on the family of the intervention. In addition, practical and emotional support once surgery has taken place is critical. We need to listen better to parents, and address their concerns, taking them with us on the journey to a decision and then continue to support them once a decision has been made and the procedure has been performed. The National Service Framework for Children sets a challenging agenda for support of children with complex medical conditions.¹² In our view this is the least parents and children should expect.

Last, but not least, we must not forget the child. Have we considered asking them for their views? Many will have significant communication difficulties or learning problems, but we should always think about the impact on them, and what they might think about being fed via a tube.

Arch Dis Child 2006;**91**:458–460.
doi: 10.1136/adc.2006.095679

Authors' affiliations

D W Vickers, Community Child Health, South Cambridgeshire PCT, Cambridge, UK
L C Maynard, East Anglia's Children's Hospices, Cambridge, UK

Correspondence to: Dr D Vickers, Consultant Community Paediatrician, Ida Darwin, Fulbourn, Cambridge CB1 5EE, UK; david.vickers@southcambs-pct.nhs.uk

Competing interests: none declared

REFERENCES

- 1 Sullivan PB, Morrice JS, Vernon-Roberts A, *et al*. Does gastrostomy tube feeding in children with cerebral palsy increase the risk of respiratory morbidity? *Arch Dis Child* 2006;**91**:478–82.
- 2 Sleight G, Brocklehurst P. Gastrostomy feeding in cerebral palsy: a systematic review. *Arch Dis Child* 2004;**89**:534–9.
- 3 Sullivan PB, Juszczak E, Bachlet AM, *et al*. Impact of gastrostomy tube feeding on the quality of life of carers of children with cerebral palsy. *Dev Med Child Neurol* 2004;**46**:796–800.
- 4 Tawfik R, Dickson A, Clarke M, *et al*. Caregivers' perceptions following gastrostomy in severely disabled children with feeding problems. *Dev Med Child Neurol* 1997;**39**:746–51.
- 5 Heaton J, Noyes J, Sloper P, *et al*. Families' experiences of caring for technology-dependent children: a temporal perspective. *Health Soc Care Community* 2005;**13**:441–50.
- 6 Kirk S, Glendinning C. Developing services to support parents caring for a technology-dependent child at home. *Child Care Health Dev* 2004;**30**:209–18.
- 7 Kirk S, Glendinning C, Callery P. Parent or nurse? The experience of being the parent of a technology-dependent child. *J Adv Nurs* 2005;**51**:456–64.
- 8 Rehm RS, Bradley JF. Normalization in families raising a child who is medically fragile/

- technology dependent and developmentally delayed. *Qual Health Res* 2005;15:807–20.
- 9 **Sleigh G.** Mothers' voice: a qualitative study on feeding children with cerebral palsy. *Child Care Health Dev* 2005;31:373–83.
- 10 **Guerriere DN, McKeever P, Llewellyn-Thomas H, et al.** Mothers' decisions about gastrostomy tube

- insertion in children: factors contributing to uncertainty. *Dev Med Child Neural* 2003;45:470–6.
- 11 **Gillon R.** Medical ethics: four principles plus attention to scope. *BMJ* 1994;309:184–8.
- 12 **National Service Framework for Children, Young People and Maternity Services.** *Disabled children and young people and those with*

complex health needs. Department of Health, September 2004 (http://www.dh.gov.uk/PublicationsAndStatistics/Publications/PublicationsPolicyAndGuidance/PublicationsPolicyAndGuidanceArticle/fs/en?CONTENT_ID=4089112&chk=0Rgaif-accessed 25 February 2006).

Endocrinology

Parsing ketotic hypoglycaemia

C A Stanley

Perspective on the paper by Bodamer *et al* (see page 483)

It has been a half century since MacQuarrie called attention to the serious issues of paediatric hypoglycaemia with his presidential address to the American Pediatric Society on “idiopathic hypoglycaemia of infancy”.¹ He pointed out that “idiopathic hypoglycaemia” could lead to seizures or permanent brain damage, and that it might have a genetic basis; however, he speculated that it was unlikely to involve insulin since insulinomas rarely occur in childhood. Over the years, there have been two types of reactions to MacQuarrie’s formulation of “idiopathic hypoglycaemia of infancy”. All too often it has been the ostrich approach of assuming that “idiopathic hypoglycaemia” is a specific entity, forgetting that “idiopathic” is the medicalese equivalent of “I dunno!”. The second type of response has been to search for underlying causes in children labelled as “idiopathic hypoglycaemia”. The latter approach has been much more rewarding, yielding an ever expanding list of new disorders, including genetic defects of hepatic glucose production (for example, deficiencies of fructose-1,6-diphosphatase, glycogen synthase, and the GLUT1 and GLUT2 transporters); a baker’s dozen defects in ketone synthesis and utilisation (for example, deficiencies of medium chain acyl-CoA dehydrogenase, plasma membrane carnitine transporter, and HMG-CoA lyase and synthase); and congenital glycosylation disorders. In addition, contrary to MacQuarrie’s speculation, many cases of “idiopathic hypoglycaemia” have turned out to have one of several genetic disorders of insulin secretion that have been recently elucidated. MacQuarrie’s original familial cases are likely to have had a dominant genetic form of hyperinsulinism. These discoveries have provided a solid foundation for rational diagnosis and management in the majority of infants and children who suffer from hypoglycaemia.

In this issue of the journal, Bodamer *et al* report studies of children with “idiopathic ketotic hypoglycaemia”.² Ketotic hypoglycaemia is, fortunately, part of a shrinking list of disorders that have been hard to rescue from the “idiopathic hypoglycaemia” wastebasket. It was originally described by two investigators from the same institution where MacQuarrie had made his observations on “idiopathic hypoglycaemia of infancy”. Colle and Ulstrom described a group of children who had recurring episodes of hypoglycaemia associated with evidence of ketonaemia and ketonuria.³ The disorder was frequently associated with suspicious past medical histories, such as low birth weight, poor weight gain, and male gender. The fact that these infants were ketotic when hypoglycaemic was appropriately highlighted as evidence that their hypoglycaemia could not be blamed on hyperinsulinism. Most cases presented between 6 months and 4 years of age, could be easily managed simply by avoiding too long an overnight period of fasting, and tended to remit by late childhood.

In studying adaptation to a standard fasting challenge of slightly more than 12 hours, Bodamer *et al* found that “ketotic hypoglycaemic” children were more hypoglycaemic, more ketonaemic, and had lower rates of glucose and amino acid turnover compared to control children. This is reminiscent of the observations by Haymond and colleagues 30 years ago that, during fasting, children with ketotic hypoglycaemia had lower levels of alanine, the major gluconeogenic amino acid, compared to control children.⁴ Both sets of data indicate that hypoglycaemia in children with ketotic hypoglycaemia reflects under-production rather than over-utilisation of glucose.

It is unfortunate that, in the title of their report, Bodamer *et al* perpetuate the

term “idiopathic hypoglycaemia” since “ketotic hypoglycaemia” is more than sufficiently mysterious and ill-defined. One would prefer that “idiopathic hypoglycaemia” would finally be excised from the lexicon of paediatrics, in a manner similar to the outdated term, “nesidioblastosis”. In particular, it is not clear whether children placed in the “ketotic hypoglycaemia” category share a common disorder or represent heterogeneous underlying disorders. Recently, Weinstein and colleagues have suggested that a significant proportion of ketotic hypoglycaemia cases may have a genetic deficiency of glycogen synthase.⁵ Since one of the co-authors of the report by Bodamer *et al*, Professor Leonard, has contributed to the advances in identifying specific defects in hypoglycaemic children over the last several decades, one hopes that the use of “idiopathic” in the title is intended to be tongue in cheek!

There are some problems with how best to interpret the observations of lower levels of gluconeogenic substrate and glucose production in children with ketotic hypoglycaemia in comparison to control children fasted for similar lengths of time. Does this reflect a qualitative or simply a quantitative difference in fasting adaptation? In the study by Bodamer *et al* and in the older studies by Haymond *et al*, ketotic hypoglycaemic children and controls are compared after similar durations of fasting rather than at equivalent physiological stages of fasting. One interpretation of these data is that children with ketotic hypoglycaemia have abbreviated fasting tolerance due to accelerated starvation. The declines in glucose production and amino acid concentrations and the activation of fatty acid oxidation and ketogenesis observed in these children may be perfectly normal, simply occurring earlier than they do in the majority of children. Similar accelerated starvation occurs in lean compared to obese individuals, in adult women compared to adult men, in children compared to adults, and in pregnant and lactating women compared to non-pregnant women. It is possible that children with ketotic hypoglycaemia merely represent the lower end of the bell shaped curve for fasting tolerance in children. Their symptomatic episodes of hypoglycaemia might simply reflect what could occur in other children if the usual overnight period of fasting were 18 instead of 12 hours.