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The right to live and the right to die

What is the minimum acceptable quality of life, asked Ian Kennedy in the Reith lectures,[1] complaining that we have no public debate about the criteria for withholding treatment from handicapped babies. This at least may no longer be true, thanks to the case of Alexandra, the baby with Down's syndrome complicated by an intestinal obstruction who the Court of Appeal decided should have a life-saving operation.

The problem with Alexandra was less clear cut than with babies severely affected by spina bifida. Professor R S Illingworth (p 612) writes of the terrible suffering of such children, and most—though not all—would agree with a policy of selective treatment at birth based on criteria, such as those of Professor John Lorber, that consider the likely quality of life.[2] Professor Lorber now urges that we should develop analogous criteria for Down's syndrome.[3] He proposes that treatment should not be withheld if the baby is simply mentally retarded, or if he or she has a simple cardiac defect; but whether to treat intestinal obstruction or major cardiac abnormalities should be for the parents to decide.

Operations to relieve intestinal obstruction are performed without question on other babies, and the implication of not treating a child who has Down's syndrome as well is that the baby may be better off dead simply because he or she has Down's syndrome. As babies with no such additional defect do live, this approach could be criticised as inconsistent. Only five months ago we pointed out that these dilemmas are a result of high-technology medicine[4]: the natural course would be for the baby to die. The existence of a treatment does not mean that it must be used in all circumstances, but where should the line be drawn? Should the baby with Down's syndrome be regarded differently from someone in the late stages of an incurable disease whose suffering would only be protracted by an antibiotic or a further operation? And is the likely degree of future suffering or unhappiness the right basis for deciding whether to treat an infant? Some of those with the most extreme defects may not suffer at all, whereas people with Down's syndrome generally have happy dispositions and on this argument would thus qualify for treatment. Moreover, some sex chromosome abnormalities, for example, may confer great unhappiness; but would anyone withhold treatment at birth? Possibly the greater difficulty of foreseeing the life of such a child and even some possibility of future treatment make the case different here. At any rate a child must be given the benefit of any doubt, as the BMA's guidance note (22 August, p 567) emphasised last week.

Alexandra's parents said that nature had "made its own arrangements to terminate a life which could not be fruitful." But do we want a society where "fruitfulness," in however wide a sense, determines worth or the right to live? Mr M J Absolon (p 611), himself the father of a teenage boy with Down's syndrome, persuasively presents the positive aspects and describes such children's enjoyment of life. But not everyone accepts that Down's syndrome is compatible with a supportable quality of life, and Professor Illingworth (p 612) questions whether the life of such a person in an institution is acceptable.

Returning, then, to the question of what is acceptable, we have to admit that for the person with Down's syndrome the answer may not always be clear. What of the parents' point of view? Apart from concern about the likely quality of life they may feel unable to cope, and indeed the wellbeing of other children and the stability of the marriage may be at stake. This, however, may be seen as a reason for having the child fostered, as is argued in our correspondence columns this week (p 611), rather than letting him die. In discussing the decision with parents paediatricians will be concerned first and foremost with the baby's interests, though they must also consider the whole family.[5] If the parents seem set to flout what the doctor believes are the baby's interests he will seek legal protection by making the child a ward of court. But should the courts make decisions in any but the most exceptional cases, such as when Jehovah's Witnesses refuse permission for a blood transfusion? A present weakness is that the guidance given to parents varies among doctors—it is likely to be pragmatic and personal, as Dr Alfred White Franklin points out (p 610); and, to go back to Kennedy,[1] the ethics have not been considered explicitly by society as a whole, which until recently has generally been content to leave such decisions with the parents and doctors. Public attitudes have changed, however—stimulated to a considerable extent by various pressure groups—and doctors must provide the most complete information about such cases and about the implications of different kinds of management, for they can no longer be left to mould society's ethics on their own. The ultimate decisions about life and death are not simply medical decisions. But to say this is not to argue that while we wait for society to set its standards the courts should be invoked. One pernicious result of the various current and threatened legal actions is the practice of more defensive medicine—for example, a greater number of operations that do not seem to be in the baby's best interests, as Professor Lorber said on *Panorama* last week.

Adult patients can make their own decisions about treatment or non-treatment, says *The Handbook of Medical Ethics*, "but

for an infant the parents must ultimately decide"—with the doctor helping the parents to understand the choices.[5] In their responses to the present case (22 August, p 567) both the BMA and the British Paediatric Association have reiterated these principles. We believe that in the absence of a clear code to which society adheres there is no justification for usurping parents' rights, or for believing that the courts are any more likely to reach a more humane solution. We must beware of that slippery slope that would lead to the nonchalant taking of lives found to be substandard, inconvenient, or expensive; but the "existence-at-all-costs" view points to a terrain no less treacherous. Letting nature take its course in certain circumstances is to acknowledge that there might sometimes be a right not to live—but we badly need to clear our confusions about what these circumstances are.

[1] Kennedy I. *Unmasking medicine*. London: George Allen and Unwin, 1981: ch 4. (Reith lectures.)
[2] Lorber J. Results of treatment of myelomeningocele: an analysis of 524 unselected cases, with special reference to possible selection for treatment. *Dev Med Child Neurol* 1971;**13**:279-303.
[3] Lorber J. Quoted by Ferriman A. Doctor tells of the babies who are allowed to die. *The Times* 1981 August 13:1.
[4] Anonymous. Withholding treatment in infancy. *Br Med J* 1981;**282**:925-6.
[5] British Medical Association. *The handbook of medical ethics*. London: BMA, 1981:32.

Pancreatic islet-acinar interactions

In current clinical practice, endocrinologists usually study and treat diseases of the pancreatic islets leaving gastroenterologists to deal with diseases of the "exocrine" pancreas. This administrative separation has, until recently, helped to obscure the functional implications of the intimate anatomical interrelation of these two parts of the human pancreas.[1] Not that islets and acinar tissue have to be anatomically associated in order to function satisfactorily: in molluscs and protochordates, for example, separate and individual endocrine cells and digestive cells are distributed in the mucosa along the alimentary tract in a "pancreatic gut," while in cyclostomes like the hagfish clumps of endocrine cells form islet organs in the wall of the bile duct but remain quite separate, anatomically, from digestive cells.[2] In mammals, however, the islets and acinar tissue have become combined within the pancreas and, indeed, apparently arise from common precursor cells.[3]

The recent recognition that the anatomical juxtaposition of islets and acinar tissue is of functional importance is based on the results of convergent studies. Animals which are spontaneously diabetic or have been rendered diabetic are known to have abnormal pancreatic exocrine function,[4] as do patients with diabetes mellitus. In rats with alloxan-induced diabetes both the synthesis and secretion of amylase are diminished but can be restored to normal by giving insulin.[5] Patients with insulin-dependent diabetes have appreciably impaired pancreatic exocrine secretory capacity[6] and morphological abnormalities of the acinar cells.[7]

In the normal pancreas the acinar tissue surrounding the pancreatic islets differs from the remaining acinar tissue by the cells being larger and having a greater content of zymogen granules, and a quite different content of enzymes, and showing both a more rapid synthesis of DNA and an increased number of mitoses.[1 8]

While the morphological basis for an intimate functional relation between pancreatic islets and acinar tissue has been recognised for a century, the intrapancreatic "portal" system of capillaries has been recognised only recently. Blood leaving the islets has now been shown to pass into the capillary meshwork within the exocrine tissue of the pancreas rather than draining directly into the pancreatic veins.[9] Studies in rabbits have shown that up to one-quarter of the total pancreatic blood flow goes to the islets and thence to the acinar capillaries before leaving the pancreas.[10] Since pancreatic venous blood contains concentrations of insulin and other hormones 20 times greater than in peripheral blood, the concentrations in the portal peri-insular circulation are presumably still higher.

The peptide hormones[11] produced by the islets influence acinar cell function. Insulin potentiates the actions of both acetylcholine[12] and cholecystokinin-pancreozymin[13] on pancreatic acinar cells, increasing both the flow of pancreatic juice and the synthesis and release of digestive enzymes, especially amylase. Specific high-affinity receptors for insulin have been shown on pancreatic acinar cells.[14] Similarly, glucagon stimulates the secretion of enzymes from the acinar cells, an effect which is potentiated by an interaction with cholecystokinin-pancreozymin and cholinergic drugs.[15] Larger doses of glucagon inhibit pancreatic secretion. Somatostatin (produced by the D cells of the islets) inhibits the function of pancreatic acinar cells,[1] as does pancreatic polypeptide, a product of the PP cells of the islets, which inhibits pancreatic exocrine secretion but also increases the synthesis of DNA in the pancreatic acinar cells.[16] Whether the insulin, glucagon, and pancreatic polypeptide which appear in pancreatic juice[17] have any regulatory function on the acinar or ductal tissues is not known. These hormones do, however, have a further wide range of indirect effects on the exocrine pancreas—for example, by modulating the intake of food. Insulin stimulates food intake,[18] and therefore (indirectly) stimulates pancreatic exocrine secretion, while pancreatic polypeptide inhibits the intake of food by its effects on the central nervous system.[19]

These interactions do not operate only in one direction, from islets to acini: important interactions probably occur among the four principal types of islet cells, since the different cell types are arranged within the islets in an ordered manner.[20] For example, somatostatin powerfully inhibits the release of insulin and glucagon; insulin inhibits the secretion of glucagon and somatostatin; while glucagon stimulates the release of the latter two hormones. Pancreatic acinar cells also have profound (albeit only indirect, so far as we know) effects on the function of the islet cells. For example, patients with exocrine pancreatic insufficiency have glucose intolerance because the islets release insufficient insulin in response to a meal, despite quite a normal capacity to do so. When the maldigestion is corrected by replacement treatment, the release of insulin and hence glucose tolerance return to normal.[21]

These indirect acinar-islet interactions depend on the "enteroinsular axis."[22] When food is broken down by the pancreatic enzymes in the intestinal lumen the pancreatic islets are stimulated to secrete hormones by the products of digestion, which elicit neural reflexes and the release of alimentary hormones from the small intestine. The most important islet-stimulant hormone is gastric inhibitory polypeptide. Gastric inhibitory polypeptide powerfully stimulates the secretion of insulin from the pancreatic islets in the presence of circulating glucose. In patients with chronic pancreatitis and exocrine insufficiency food is not satisfactorily digested; release of gastric inhibitory polypeptide from the small intestine by the breakdown products of fat and carbohydrate is therefore less than normal; and release of insulin is therefore