The fundamental problem lies in the apparatus itself; equipment currently available for the use of the handicapped is poorly designed and often unsuited to the child's needs. (In the Wolfson Centre series, 19 out of the 31 appropriately chosen chairs required structural modification.) Much of the equipment is traditional in concept and style, taking no account of modern technology, anthropometry, or ergonomics, being designed mainly by engineers with no clinical contact with the patients involved. The designer of apparatus for this specialized group of patients must apply much stricter criteria to his data collection and design method.¹ In design terms, being handicapped means that the patient has a reduced flexibility and cannot adjust to poor ergonomics as can a normal child. A design must be based on the problem reduced to its simplest form. The concept of a "chair" is irrelevant; it is the postural requirements of the patient that the designer must attempt to satisfy.

The second major fault of currently available apparatus is its complexity. Most chairs are multivariable and can be adjusted to anatomically and ergonomically nonsensical positions. True flexibility can be achieved only from the concept of a dynamic environment in which the child can develop. Simple adjustments which always maintain correct posture reduce the inappropriate use of chairs. Fewer structural modifications are required, which eliminates some of the delays in delivery. In the use of the Cell Barnes chair,² which is purpose-designed for the severely handicapped, no structural modifications have been required in over four years' use in a clinic seeing about 100 new patients per year. Most children can be appropriately equipped with the standard chair; necessary modifications are minor and can be achieved immediately in the clinic with use of direct moulding techniques with plasazote or similar materials to provide individual, specifically corrective forms. Adjustments can be made at each visit with the improvement in the child's posture and control.³

With the present Department of Health tendering system, manufacturers are reluctant to spend money on the development of ergonomically designed equipment which departs from conventional design for the handicapped as they may not receive a contract and would be unable to recoup development costs. Until attention is drawn to the positive harm done and the waste of resources resulting from the unsuitable and misapplied use of apparatus, and until adequate incentives are offered to designers and manufacturers, this regrettable state of affairs is likely to continue.—I am, etc.,

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- ¹ Taylor, R. H., *Community Health*, 1972, 3, 162.
- ² Green, E. A., and Taylor, R. H., *Physiotherapy*, 1969, 55, 376.
- ³ Green, E. A., and Taylor, R. H., *Physiotherapy*, 1971, 57, 68.

Tragic Dilemma

SIR,—I am a doctor and have been the parent of a child handicapped similarly to the one with which your leading article "Tragic Dilemma" (9 December, p. 567) is concerned.

When I first noticed—and it took several

months for the condition to become apparent—that my child was suffering from something so serious, the world went dark around me. And after the tests proved that an operation might offer some hope, I experienced almost a triumph: something could be tried for him. Those were the days of the first neurosurgical efforts on such conditions and, inevitably, each patient had to serve as a guinea-pig as well.

I remember the five times I held my son's hand as he was directed towards the operating theatre and left him on the trolley to pass through the swing doors. I remember him returning to the ward, going through the suffering of the first postoperative days, undergoing investigation after investigation, receiving one injection after the other. I have always been fully aware of the "physical" tragedy his life was until his death a few years later. If left alone, he would have certainly died much sooner. By going on with whatever medical treatment was available then we were only prolonging his day-to-day suffering. But this suffering was definitely reduced and there was the belief that perhaps, were he to survive, the quality of his life was being improved. I have never regretted what I did then.

Some years later I worked in a hospital where a little girl with my child's illness was treated successfully. Recent advances and knowledge based on experience had made her capable of running around, chattering happily, developing like any other normal child, enjoying life—herself and her parents. Then, even more than ever before, I felt as a parent and a doctor that my child's life and death were, after all, justified.—I am etc.,

MEDICAL PARENT

Anaesthesia in Sickle-cell States

SIR,—We have read the paper by Professor K. A. Oduro and Dr. J. F. Searle (9 December, p. 596) entitled "Anaesthesia in Sickle-cell States: A Plea for Simplicity" with interest. The insertion of the ambiguous word "simplicity" into the title makes it possible that anaesthetists might interpret the article as a reassurance that the anaesthetic management of sickle-cell states requires no particular skill. This would be regrettable because, on reading the text, one becomes aware that the authors are in fact advocating something very different. They recommend a painstaking preoperative preparation, a most carefully administered anaesthetic, and efficient postoperative care which includes postoperative oxygen for 12–24 hours.

Despite careful anaesthetic management, there were six postoperative deaths in the 505 patients in the series. Although sickling was thought to have played some part in the death of only two of these patients, one cannot categorically exclude it as a contributory factor in the other four. It was difficult to relate anaesthetic death and sickle-cell haemoglobin from the figures given in the article because there were no data on a comparable series of anaesthesia in non-sickling patients.

With their great experience in this field Professor Oduro and Dr. Searle believe that "a simple anaesthetic technique together with good postoperative care can provide safe general anaesthesia for patients with sickle-cell states" (our italics). In a recent review¹ we concluded that "even if every

precaution is taken, anaesthesia for a patient with sickle-cell disease may prove a hazardous and at times a fatal undertaking." This apparent difference in opinion is at least in part explainable because the vast majority of patients with sickle-cell haemoglobin (that is, with a sickle-cell state) are sickle-cell trait carriers. It is generally agreed that the anaesthetic risk for the sickle-cell trait carrier during routine surgical procedures must be extremely small. In contrast the anaesthetic risk for the small minority of patients with sickle-cell haemoglobin who have sickle-cell disease (that is, sickle-cell anaemia, sickle-cell haemoglobin C disease, and sickle-cell thalassaemia) will be very much higher. For example, there were only 42 patients in Professor Oduro and Dr. Searle's series known to be in this latter category and one of them, a 22-year-old woman (having a McMurray osteotomy) died six hours post-operatively with a "sickle-cell crisis."

In order to minimize the anaesthetic hazards, even in the emergency situation, the anaesthetist in Britain should attempt to divide all patients who have been shown to possess sickle-cell haemoglobin into the low-anaesthetic-risk sickle-cell trait carrier and the high-anaesthetic risk patient with sickle-cell disease. It is fortunate that this differentiation can usually be achieved by simple laboratory tests such as a solubility test to recognize sickle-cell haemoglobin combined with a reticulocyte count. The exact diagnosis of a patient provisionally labelled as having "sickle-cell disease" can be arrived at the next day using more refined techniques such as electrophoresis. The information that the patient has sickle-cell disease should result in a reappraisal of both the diagnosis and the need for operative interference. Even if a careful general anaesthetic is subsequently administered to a patient with sickle-cell disease, it would be unwise for the anaesthetist to imagine that this will always prove to be a safe procedure.—We are, etc.,

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- ¹ Howells, T. H., Huntsman, R. G., Boys, J. E., and Mahmood, A., *British Journal of Anaesthesia*, 1972, 44, 975.

Sarcoid Heart Disease

SIR,—Your recent leading article (16 December, p. 627) draws attention to our report¹ of six new cases of fatal myocardial sarcoidosis. Since that report was completed, from further inquiries I have collected a total of 44 additional new cases in the United Kingdom. My inquiry was prompted by the fact that I had seen, during life, three of the six patients in our original report and, while I was aware of the existence of sarcoid involvement of the myocardium, I had failed to make the aetiological diagnosis. This suggested to me that the condition was more common than is recognized, and the reaction of many colleagues confirms this impression. They have neither made the diagnosis nor thought of its possibility.

Of the total of 50 patients, 17 are from the East Anglian area and these have all been seen by me personally. Most of the other cases come from a relatively small number of centres and this suggests that the diagnosis is overlooked elsewhere. While all