MYASTHENIA GRAVIS IN A 2-YEAR-OLD TREATED BY THYMECTOMY

G. J. SUTIN, M.B., CH.B., M.R.C.P. (EDIN.), D.C.H. R.C.P. & S. (ENG.), *Part-time Paediatrician*, AND R. P. HEWITSON, F.R.C.S., F.R.C.S. (EDIN.), *Part-time Thoracic Surgeon, Somerset Hospital and Red Cross War Memorial Children's Hospital, Cape Town*

Myasthenia gravis is a rare illness in childhood. The usual approach to therapy is conservative and there are only few instances documented where thymectomy has been undertaken.[1,2] This report concerns an infant, who may be the youngest patient on record, in whom progression of the disease justified this radical procedure.

CASE HISTORY

M.P., a Coloured female infant, born on 15 November 1963, was first seen by one of us (G.J.S.) on 24 September 1965. About 3 months before this visit her parents had noted drooping of the left upper lid, which they spontaneously volunteered had become more marked as the day progressed. A few days before being seen the right eye had become similarly involved. She had also shown undue lethargy towards afternoon, and was taking to bed much earlier than usual. On physical examination she had a rather expressionless face, and bilateral ptosis of a marked degree which she was unable to correct. The knee reflexes were probably diminished, but there was no other evidence of neurological deficit. She was admitted to the Somerset Hospital where a dramatic response to 0·5 mg. of prostigmine intramuscularly, confirmed the diagnosis of myasthenia gravis.

A satisfactory response was obtained to prostigmine, 7·5 mg. orally twice a day, and she was dicharged on 15 October 1965. The effect, however, gradually wore off, and when seen on the morning of 13 December 1965, she was found to have a partial bilateral external ophthalmoplegia, which by afternoon had become total, when seen by Dr. L. Schrire.

She was admitted to the Red Cross War Memorial Children's Hospital, Rondebosch, on 15 December 1965. Increased dosage produced a variable and inconstant response, and the impression of peripheral muscular involvement became more dominant. Thymectomy was performed on 29 December (R.P.H.). The thymus removed *in toto* was macroscopically normal, with no evidence of neoplasia, lymphoid follicles or germinal centres found on histology (Dr. C. E. Watson).

Following operation, improvement was rapid and progressive. After 48 hours, markedly increased responsive-

ness was noted particularly in the movement of the upper lids. She was discharged on 20 January 1966 on prostigmine, 2·5 mg. *b.d.*

When seen again in the company of her parents, on 7 February 1966, they enthused about the remarkable change in her and commented particularly on her 'radiance', increased activity, and improvement in speech. She had a slight divergent squint, but otherwise nothing of note. She had been off prostigmine for 36 hours, and this has not been reintroduced since.

When last seen on 30 May 1966, she was doing very well. The strabismus was still in evidence, and a transient torticollis was attributed to a non-myasthenic cause. In a subsequent telephonic communication (14 August) the mother's enthusiasm was unabated, and she stated that the squint was no longer apparent to them.

The accompanying photographs show the patient immediately pre-operatively on 29 December 1965 (Fig. 1) and postoperatively on 6 January 1966 (Fig. 2). These were taken at approximately the same time of day before the morning dose of prostigmine had been given.

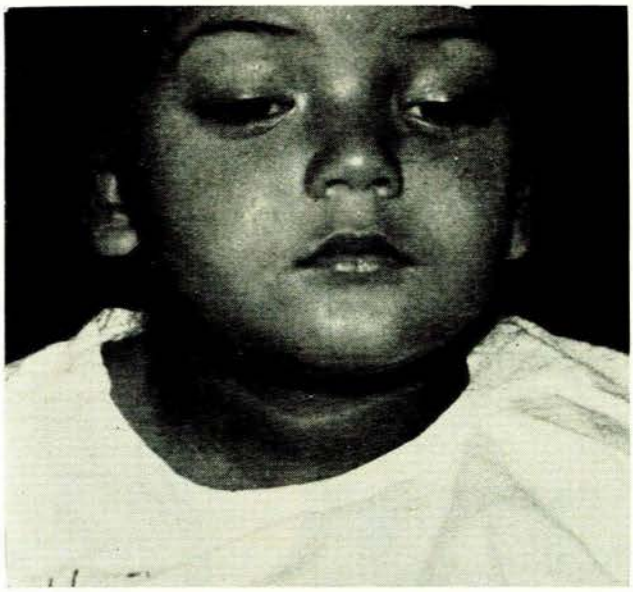

Fig. 1(a)

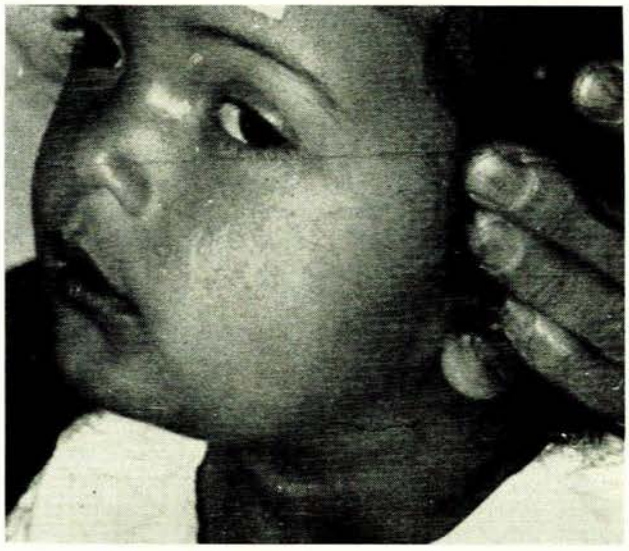

Fig. 1(b)

Fig. 1. Immediately before the operation on 29 December 1966. No morning dose of prostigmine had been given. *1(a).* The bilateral ptosis and facial immobility can be seen. *1(b).* Strabismus clearly shown.

Fig. 2. Postoperatively on 6 January 1966. The morning dose of prostigmine had not yet been given. Note absence of ptosis, alert facial expression and free mobility of the eyes. On looking forward, divergent strabismus present.

DISCUSSION

It has been repeatedly stressed that in myasthenia gravis the younger the patient and the shorter the history, the better the response to thymectomy.[1,3,4] It has also been stated that a stable and complete remission is more likely, following successful surgery.[1,5] Further, once external ophthalmoplegia develops, prostigmine improves the ocular palsies only slightly, if at all;[6,7] and the longer these are allowed to persist the greater the likelihood of subsequent operative correction being necessary.[7] These criteria, along with the progressive deterioration, influenced the decision towards surgery. The subsequent course has been most gratifying. To date, in addition, she has shown no signs of increased liability to infection, nor has her growth and development been affected in any adverse manner.

Keynes[2] states that 'two moribund infants aged 2 and 4 years have been almost literally snatched from the grave by operation', but supplies no further details. We know of no other report of patients in the very young age-group who have been treated in this manner.

SUMMARY

A case is reported of thymectomy performed on a toddler 25 months of age, with myasthenia gravis. Pre-operatively the patient had shown a progressively deteriorating clinical course, and the result to date has been most gratifying. The literature indicates that there is a good chance that the remission will be complete and permanent. She is one of the youngest, if not the youngest, patient to have been treated in this manner.

We wish to thank our medical and nursing colleagues at Somerset Hospital and the Red Cross War Memorial Children's Hospital for their interest and help; Dr. L. Schrire for his valuable assistance; Profs. M. Luntz, F. Ford, and F. Forman for their valuable advice and guidance; Dr. G. Fisher for the pre-operative and Mr. B. Todd for the postoperative photographs; and Drs. R. Nurok and J. F. W. Mostert, the respective Medical Superintendents, for permission to publish.

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