Post-operative Mental Development in Patients with Hydrocephalus and Craniosynostosis

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Studies on children treated for craniosynostosis and hydrocephalus have focussed mainly on medical aspects in the past. However, in recent years the intellectual and social development of such children are also being studied. These children suffer from rejection since birth, due to obvious gross craniofacial deformity, brain anomalies and mental retardation(l). The intellectual deterioration is mainly caused by raised intracranial pressure with or without hydrocephalus and/or craniofacial syndrome. Psychosocial assessment is important to determine suitability for surgery, providing pre- and post-operative counselling, long-term psychosocial development and rehabilitation. The importance of longitudinal studies in the postoperative management of these patients cannot, therefore, be ignored. In order to evaluate the degree of mental retardation, and explore the factors, including interval between operation and testing, type of deformity and age at operation, affecting mental development we have assessed 350 surgically treated patients of craniosynostosis and hydrocephalus.

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Material and Methods

We studied 50 surgically treated patients of craniosynostosis, and 300 patients of hydrocephalus attending the craniosynostosis and Hydrocephaus Clinic of Department of Pediatric Surgery at the All India Institute of Medical Sciences. For the purpose of analysis, patients with craniosynostosis were further grouped into simple and complex type, the latter were associated with hydrocephalus and/or craniofacial syndromes. Patients who had not undergone surgery or those who had no visual acuity *i.e.,* no perception to light were excluded.

Psychological assessment was done using the Vineland Social Maturity Scale(2), to assess the social maturity of the child and to assess self help skills. The AIIMS developmental scheduled) was prepared to assess the mental performance in terms of quotient (MPQ), of children from 0 to 5 years of age and to calculate the motor, adaptive, language and personal social quotient separately. Binet Kamath test(4) was used for children above three years of age to assess the intelligence quotient (IQ). The Seguin form board(5) was used to know the concept formation of the child and to have a rough estimate of IQ in uncooperative children. The assessment were made preoperatively, at 1 month, 3 months, 6 months and at yearly-intervals following surgery. The term MPQ was used for the Mental Performance Quotient of children below 5

years of age assessed on AIIMS developmental schedule.

Results

The preoperalive mean MPQ/IQ of hydrocephalic children was 43.5 ± 25.4 and those of craniosynostosis was 85.5 ± 17.9 *(Table I).* Amongst the latter group, mental performance in simple craniosynostosis, *i.e.,* craniosynostosis without hydrocephalus and craniofacial syndrome was 92.6 ± 16.6 and 97.2 ± 13.4, respectively. The mental performance in complex craniosynostosis, *i.e.,* craniosynostosis with hydrocephalus and craniofacial syndrome was 75.6 ± 28.9 and 81.8 ± 15.5, respectively *(Table II).* Postoperative follow-up studies of mental assessment, in both the groups, revealed an increase in MPQ after one month of surgery *(Table I).*

A comparative post-operative performance in the two groups revealed 54% of children with craniosynostosis had an average intelligence, *i.e.,* MPQ/IQ (>90) as compared to 18% of hydrocephalic children.

Study of pre and post-operative MPQ according to age at operation revealed increase in MPQ in those operated at younger age, *i.e.,* (<3 mo). The MPQ was 100 ± 9.0 in craniosynostosis and 66.8 ± 27.5 in hydrocephalus whereas in cases operated above 2 years of age, MPQ was 90.5 ± 13.6 and 44.9 ± 23.6, respectively.

Discussion

In the last decade, the mortality and morbidity amongst hydrocephalic and craniosynostosis patients has considerably declined with appropriate surgical therapy.

TABLE I–*Follow up of Mental Performance*

Follow up interval	Craniosynostosis			Hydrocephalus		
	N	Mean	SD	N	Mean	SD
Pre-operative	41	85.5	17.9	180	43.5	25.4
Post-operative						
< 1 mo	8	79.5	15.6	114	44.6	23.6 NS
1- 3 mo	17	88.9	12.1	131	51.3	28.8**
3- 6 mo	24	89.7	14.7	97	58.5	24.5***
6-12 mo	24	89.8	14.5	99	55.4	24.7***
1- 2 yrs				101	57.4	25.7***
2- 3 yrs	26	88.2	15.2			
≥ 3 yrs	13	95.3	16.2			
Highest MPQ				298	60.7	29.5

MPQ : Mental performance quotient.
NS : Not significant.
** : Indicates significance at 0.01 level.
*** : Indicates significance at 0.001 level.

TABLE II–*Mental Performance of Craniosynostosis With and Without Associated Syndrome and Hydrocephalus*

		Post operative Mean MPQ/IQ	SD
A	Cranio. without hydrocephalus	92.6	16.6
	Cranio. without syndrome	97.2	13.4
B	Cranio. with hydrocephalus	75.6	28.9
	Cranio. with syndrome	81.8	15.5

A = Simple craniosynostosis;
B = Complex craniosynostosis.

The ultimate aim of surgical treatment has been to achieve maximum brain potentials. Psychosocial assessment has played important role in evaluation of behavior problems and mental performance and has thereby helped in early prevention of complications of the disease. Most studies have revealed that the risk of mental retardation exists in all forms of craniosynostosis and hydrocephalus(6,7). However/ the severity of retardation may vary. considerably depending on degree of raised intracranial pressure, associated brain damage, anomalies and syndromes, age at presentation and at surgery, degree of visual loss and status of motor function. Dennis *et al.* (8), reported that the outcome of early hydrocephalus is an uneven growth of intelligence during childhood. They found that the origin of the cognitive deficit was neither in the hydrocephalic condition itself nor in its treatment, but rather in the development of brain anomalies and symptoms to which the hydrocephalic child is prone. Upadhyaya *et al.(7)* in their long term study of shunt treated hydrocephalic children reported that these children improve progressively upto 5 years. The incidence of mental retardation reported for simple craniosynostosis is lower than for the complex syndromes. This may be the result of progressive rise in intracranial tension due to craniosynostosis and/or associated hydrocephalus or genetic factors related to syndromes(6). On autopsy, Gross(9) has shown important structural changes in brain of patients with Crouzon's and Apert syndrome. These facts suggest that mental retardation is related not only to raised intracranial tension but also to the structural changes in the brain. In Apert syndrome, physical disabilities also contribute to retardation. The present study revealed that preoperative MPQ/IQ of craniosynostosis patients was in dull normal (70-<90) category as compared to children with hydrocephalus, who were in the category of mild retardation (50-<70). Further, it was observed that simple craniosynostosis patients had an average MPQ/IQ whereas complex craniosynostosis associated with hydrodephalus and syndromes were dull normal. These observations suggested a definite correlation between mental retardation and severity of craniosynostosis as against other reports(10,ll) which observed poor correlation. Raimondi(12) and Tromp(13) reported that the incidence and degree of mental retardation in such patients depended on the degree of preoperative irreversible brain damage, age at surgery, nature and severity of illness. The present study also revealed that all the patients of both the groups showed initial fall in MPQ in the first month of post-operative period and then there was progressive rise with age. Also, younger the patient at surgery, better was the post-operative MPQ. Older hyprocephalic patients were more often retarded but it was not always true with

craniosynostosis. However, inspite of some improvement, majority (60%) of the hydrocephalic subjects remained retarded with highest MPQ/IQ (60.7 ± 29.5) as compared to craniosynostosis [10%, mean MPQ/IQ (95.3 ± 16.2)]. Psychological problems such as anxiety, helplessness in these children were reported by the parents. Also, the emotional disturbances of the parents, like fear of development and struggle to raise such a child was noticed, as also reported by others(14,15). Since these patients are not only mentally retarded but also handicapped because of associated physical disabilities, management requires much more attention and careful observation than other groups of mentally retarded children. Behavior modification and parental counselling are the best mode of therapy for any associated behavioral problem and for anxious parents(7,16). However, frequent psychosocial assessment is essential to evaluate the comparative mental development with growth and passage of time.

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