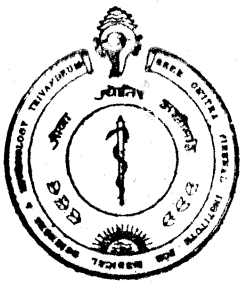

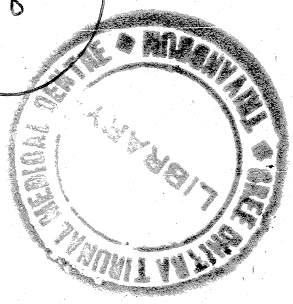

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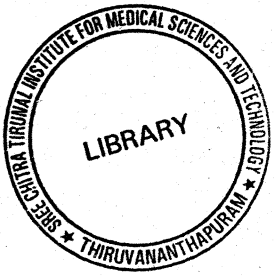

LIST OF PROCEDURES DONE
PROJECT REPORT

NAME : ALEVOOR RAJARAM BHAT

PROGRAMME : D.M. NEUROLOGY

MONTH & YEAR OF SUBMISSION : Oct. 1994

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LIST OF PROCEDURES DONE
PROJECT REPORT

TITLE OF THE PROJECT

1. SUBACUTE SCLEROSING PANENCEPHALITIS (SSPE):EXPERIENCE OF A TERTIARY REFERRAL CENTRE IN KERALA, S.INDIA

2. IDIOPATHIC INTRACRANIAL HYPERTENSION: A RETROSPECTIVE STUDY

NAME ALEVOOR RAJARAM BHAT

PROGRAMME D.M. NEUROLOGY

MONTH & YEAR OF SUBMISSION 1994

SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES & TECHNOLOGY THIRUVANANTHAPURAM-695 011

Name	
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Date	

CERTIFICATE

I, Dr ALEVOOR RAJARAM BHAT.....hereby declare that I have actually performed all the procedures listed / carried out the project under report.

Signature.....

Place : TRIVANDRUM

Name in ALEVOOR RAJARAM BHAT.....

Date : 19.10.94 capital letters

Forwarded. He has carried out the minimum requirement of procedures / etc.

Signature

Head of the department

CONTENTS

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Date: 19/10/94
Thiruvananthapuram.

Alevoor Rajaram Bhat

Subacute Sclerosing Panencephalitis (SSPE) : Experience of a Tertiary Referral Centre in Kerala, S. India

Introduction

Historic Review of SSPE

After the abatement of epidemic of encephalitis lethargica in the early part of this century a sporadic encephalitis with a subacute course was recognized. In the first description of SSPE the white matter component of the disorder was not fully appreciated. In 1950 it was known as Dawson's inclusion body encephalitis. Later it was termed subacute sclerosing leuco encephalitis. By 1969, the name subacute sclerosing panencephalitis was universally accepted. In 1967, a measles like virus was believed to be associated with SSPE which was confirmed[1]. Measles protein identified as M-protein has been implicated in pathogenesis of SSPE.

Epidemiology:- Epidemiological investigations have been key in the development of an understanding of this disease and have been brought about through activities of the USA National SSPE Registry[1]. SSPE is world wide disease with an

incidence rate about 1 per million children a year[2]. In US the disease in more common in the southern states and the Ohio River Valley. The incidence of measles in India is 190 per 100,000 population and the incidence of SSPE is reported to be 7 per 100,000 natural measles infection which is reduced by measles vaccination to 1 per 100,000 vaccination[3]. Saha et al[4] have reported high incidence of SSPE in South India i.e. 21 per million population. Miller et al[5] reported risk of SSPE following measles in England 4/100000 cases and risk after vaccination as 0.14/100,000 cases.Vaccination has lowered the incidence of SSPE by 5-50 times[2].

Approximately one half of the persons had measles before age 2, 70-80 per cent before age 4. Males outnumber females by 2.3-1 and patients commonly come from large families and from rural rather than urban areas[1,2,5,6,7,8]. The mean incubation period is 6-7 years with 85% cases occurring between 5 and 14 years[6]. Shafyi[9] et al in their study found interval between measles infection and onset of SSPE ranging for 2-22 years with mean of 8.8 years. Detels et al reported that measles occurred at median age of 15 months in SSPE and at 48 months in control[8].

Racial and ethnic factors

Possible genetic factors are indicated by increased incidence of the disease in various ethnic groups. In Israel, the incidence of SSPE is sixfold higher in Arabs and Sephardic Jews than in Ashkenazic Jews and in South Africa it is higher in coloured persons[2].

Head Injury : The association of SSPE, with head injury has been noted in a number of case reports and a controlled epidemiologic study. Similarly exposure to birds is significantly more common is patients with SSPE[11].

Pathogenesis and Pathology:-

One half to one third of otherwise uncomplicated patients with measles have virus involvement of CNS as indicated by transient electroencephalographic changes and CSF pleocytosis. It is likely that patients with SSPE have CNS seeding by measles at the time of primary infection. In majority virus is cleared by immune mechanism shortly after primary measles[1]. Cellular rather than humoral immunity is required both for recovery from natural measles[1]. Patients

with SSPE do not have enhanced cell mediated immunity to measles virus compared with controls. The viruses isolated from their brains are defective compared with wild type measles virus. Data from experimental studies suggest that a virus mutation that restricts the expression of matrix protein (M Protein), is responsible for SSPE. Virus mutation and antigenic modulation are the currently proposed inducers of the defect that characterizes the SSPE virus[2].

Pathology

Pathologic changes are found both in gray and white matter and tend to be more pronounced in occipital than in frontal hemisphere. Histologically foci of demyelination are seen and gliosis and proliferation of microglia may be pronounced. Measles virus antigen can be found in neurons and glia even in absence of inclusions.

Clinical features:

Natural History:- Typical patient with SSPE has a history of primary measles infection at an unusually young age, usually under 2 years of age followed by asymptomatic

period of 6-8 years. The onset of the disease is typically evidenced by a decline in the child's scholastic performance followed by progression through characteristic stages[2,6].

Stage I- Mental and behaviourial changes, forgetfulness, irritability and lethargy.

Stage II- Myoclonic jerk, Dyskinesia, athetoid postures, ataxia.

Stage III Decerebrate, Decorticate, Coma

Stage IV- Loss of cortical functions - Flexion posturing of limbs.

The disease begins insidiously. Clinical disease may begin acutely with hemiplegia or seizures. Death typically occurs in 1-3 years. In a large Lebanese series 95% patients died in less than 6 years. Those 5% patients who survived more than 6 years represent a distinct subgroup[12]. US registry data indicate a median length of survival of about 1 year, a 5-year survival of 10 percent and essentially all patients dead by 10 years. Trends changed after 1980, sex

ratio is gradually decreasing. Age of onset of neurological symptom has gradually increased and is now close to 14 years. Average measles/SSPE interval has increased to 10.5 years[1]. Further, apart from classical SSPE, other presentation of SSPE - a chronic form, a "fulminant" form leading to death in weeks to months and a stuttering type like multiple sclerosis have been recognised[13].

Visual symptom occur in about half the cases and are usually due to a focal chorioretinitis, although cortical blindness also occurs[14]. Reported spasm or myoclonic jerks occur in nearly all cases. These spasms occur with rapid onset but with a delayed or 'hung-up' relaxation phase that lasts 1 second. These repetitions in SSPE is reflection of the periodic complexes seen in EEG of these patients. This stage II lasts for 3-12 months. As the disease progresses to stage III patients become bedridden due to choreoathetosis, dystonia and rigidity often with extensor posturing. This stage lasts for 3 months to 18 months. Stage IV patients develop flexor posturing. Dyken et al[15] showed that there is correlation between staging and neurological disability, stage I corresponds to 0-30% disability, stage II with 31-55 percent disability, Stage III with 55-80 percent disability and stage IV with 81 to 100 percent disability.

Laboratory findings

CT scan is normal early in the disease with progressive appearance of enlarged ventricles and widened sulci indicating cerebral atrophy and multiple low density lesions probably corresponding to areas of demyelination and encephalitis. There is no correlation of disease stage duration with CT abnormalities[2]. EEG may be normal in the early stage. Characteristic periodic complexes occur in nearly every patient with SSPE in at least at some stage of the illness. The complexes consists of high voltage (300-1500 uv) repetitive polyphasic and sharp slow wave complexes ranging from 0.5-2 sec. in duration and recurring every 4-15 seconds[16]. The following characteristics of periodic complexes (PC) have been outlined. (1) They are bilateral usually synchronous and symmetrical (2) they are stereotyped (3) they usually contain two or more delta waves (4) The amplitude is usually around 500 uv (5) they recur with a fair regularity every 4-5 sec. When clinical myoclonus are present there is one to one relationship between jerks and PC[18]. In addition to periodic complexes other abnormalities seen are polymorphic delta activity, electrodecremental response following PC and various types

of epileptic form discharges[18]. Basic rhythm becomes disorganised and slower and the interval between the periodic complexes may shorten as the disease progresses.

CSF examination in SSPE shows no abnormalities in pressure, cell count, total protein or sugar. Elevation of measles antibody titre and of IgG concentration in the CSF is seen in all patients. As the disease progresses, protein may be elevated. In a number of studies, IgG concentration in CSF was between 10-54 mg/100 ml as compared with 5-10 mg/100 ml in normal children. More specific than IgG concentration is the fact that 50-80% of the IgG in CSF is specific antibody against measles virus protein. Among the methods commonly used are complement fixation (CF), hemagglutination inhibition (HAI) and virus neutralization (VN), Highly sensitive Elisa assay are useful in detecting virus-specific IgM as well as IgG. Elevated measles antibody titre in the CSF is more diagnostically significant. They range from 1:4 to over 1:512 in both CF and HAI tests. These CSF antibodies are synthesized intrathecally[6]. Brain biopsy and histopathology may reveal any one of the characteristic features including neuronal and glial cell inclusions, subacute inflammatory vascular changes, subacute demyelination and extensive gliosis. The

relationship between measles virus and SSPE has been firmly established and measles virus has been isolated from cultured brain cells of patient with disease[17].

Treatment : Several experimental antiviral and/or immunomodulatory therapies have been tested in SSPE patients. The most extensively tested agent is Isoprinosine which has both antiviral and immunomodulatory properties. Dyken et al[14] reported stabilization in two-thirds of patients while other studies conclude that drug is ineffective[17]. Interferon has been tested in a number of small scale clinical trials. Interferon also has both antiviral and immunomodulatory properties. Natural alpha interferon has been used in most trials.

Prognosis: SSPE is fatal in over 95% of cases, 80% individuals succumbing between 9 months and 3 yrs. Stabilization or partial remission can occur at any stage of the disease, most frequently in stage 2 or 3[6].

Aims of the Study

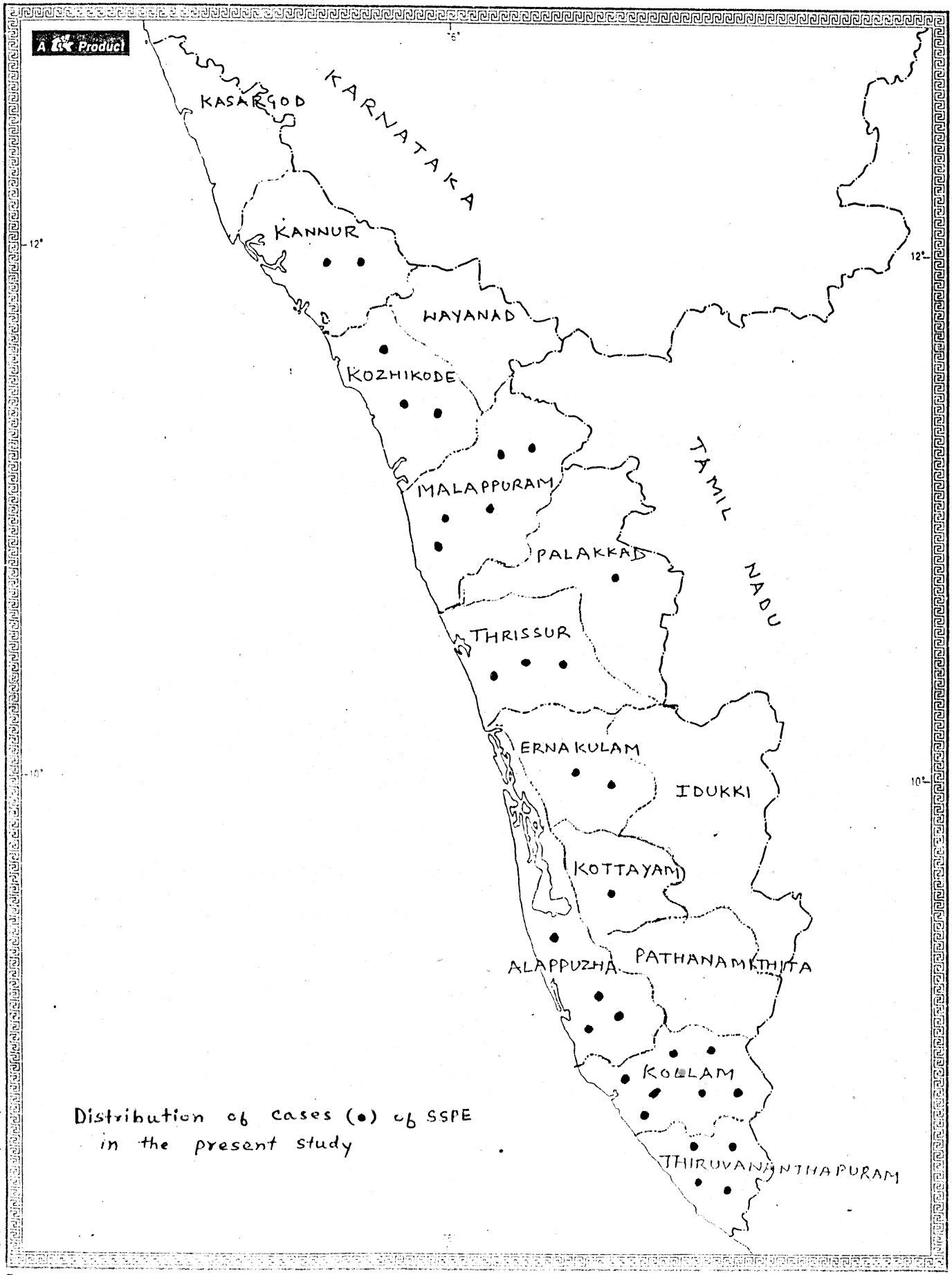
1. To determine age of onset of measles in patients of SSPE.
2. To determine interval from onset of measles to SSPE.
3. To study clinical characteristics of patients with SSPE.

Materials & Methods : 32 patients of SSPE were diagnosed from Jan. 1984 to July 1992. Case records of these patients were studied in detail. SSPE was diagnosed based on Dyken's criteria[1] i.e. patient fulfilling any three of following criteria-

1. Typical clinical presentation.
2. Typical EEG. pattern i.e. stereotyped periodic complexes.
3. Typical histologic findings either by brain biopsy during life or postmortem studies.
4. Hyperglobulinorrhachia greater than 20% of total protein.
5. Elevated serum and cerebrospinal fluid (CSF) measles antibodies, titres $\geq$ 1/4.

EEG was recorded on an eight channel machine as per standard protocol. Measles antibody titres were measured by ELISA or Hemagglutination inhibition test. Staging of the SSPE was done as per modified Jabbour criteria[6]. Postal follow up was done using a questionnaire to assess the neurological status of the patients.

KERALA – OUTLINE



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The territorial waters of India extend into the sea to a distance of twelve nautical miles measured from the appropriate base line

Results

The study consisted of 32 patients with age ranging from 2.5 to 19 years (Mean 9.78 years, median 11 years). There were 24 males with age of 2.5 to 16 years (mean 9.37 years, median 10 years) and 8 females in 5-19 years range (mean 11 years, median 10 years). Male:female ratio was 3:1. (Age distribution is shown in table 1,2). Most of the patients came from a rural background (rural:urban ratio was 7:1). History of prior measles was obtained in 19 patients (59.3%) (Table 3A). Measles occurred between 1-8 years of age. (mean age of onset of measles was 2.56 years and median 2 years). Age range in this group was 2.5-14 years. (mean - 9.05 years, median 9.5 years). Measles to SSPE interval ranged from 1-12.7 years (Table 3B). (mean 6.10 years, median 6.5 years), mean interval for males and females were 6.46 and 4.75 years respectively. In this group male:female ratio was 3.75:1. Mean duration of symptoms at onset was 4.8 months (range 1-12 months).

The second group who did not give history of prior measles consisted of 13 patients (9 males and 4 females).

The mean age in this group was 11.69 years and median 11 years (range 6-19 years). Mean duration at presentation was 6.92 months. The first group (with h/o measles) differed from the other in the following respects. (1) age of onset was earlier (2) male:female ratio was higher (3) mean duration of symptoms was shorter (4) the progress of the disease was faster. In measles group, 57.8% patients were in stage II and 21.05% in stage III within 6 months of onset as compared to 38.37% (Stage II) and none (Stage III) in other group.

Cognitive decline and myoclonic jerks were the commonest presenting features (93.7% and 84.3%) (Table 4).

In 11 patients (34.3%) CSF IgG was very high (>50% of the total CSf protein) (Table 5). Very high CSF measles antibody titres (>1:1000) were present in 5 patients (Table 6), 4 of them had history of prior measles. There was no corelation between CSF measles antibody titres and CSF IgG. 19 patients underwent CT scan of brain. In 3 there were white matter hypodense lesions while the rest were normal. EEG - All the patients had periodic complexes in their EEG. 20 had slow wave PC rest had sharp wave PC. In 3 patients P.C. were brought out by I/v diazepam. In 6 patients burst

suppression was present. Postal follow up responses were obtained in 12 cases (37.5%). 2 patients (No.10 and 12) died after 1 and 5 months of onset respectively. 3 patients improved to independent existence (No 1, 6, 20) after 1 to 3 years. Other 7 patients were bed bound after 1-5 years of onset of disease (Table 7). 6 out of 7 were males, 3 of them gave history of measles in the past. There were 2 patients each with disease duration of 2, 3 and 6 months respectively while one had 9 month disease duration at diagnosis. 6 patients had CSF IgG more than 30% of CSF proteins while only one had high CSF antibody titre (1:1000).

Table 1

Age distribution & SSPE

	Age Group	No.	%
1	1 - 4	2	6.25
2	5 - 9	12	37.5
3	10 - 14	15	46.8
4	15 - 20	3	9.37
Total		32	

Table 2

Demographic Characteristics of patients

Age: range 2.5-19 yrs.	Mean 9.78 median:11 yrs
Ethnic composition:	Hindu 48.4% Muslim 24.2% Christian 27.2%
Economic Status	: A(43.7%), B1(25%), B(12.5%), C(12.5%), D(6.2%)
Rural:Urban = 7:1	
Male : female	: 3:1
Age distribution (females)	:range 5-19 years mean 11 yr median 10 yr
Age distribution (males)	:range 2.5-16 yr mean 9.37 yrs median 10 yr.

A = Income ≤ Rs.300 per month
B1 = Income Rs.301 - 500/month
B = Income Rs.501 - 1000/month
C = Income Rs.1001 - 1500/month
D = Income > Rs.1500/month

Table 3A

H/o measles	: Yes 59.3%
Onset of measles	:Age range 1-8 yrs mean 2.56yr median 2yr.
Measles-SSPE interval	:range 1-12.7 mean 6.10yr median 6.5 yr.
Age of patient (with h/o measles)	:range 2.5-14 mean 9.05 yr median 9.5 yr.
Age of the patient (without h/o measles)	:range 6-19yr mean 11.69yr median 11yr.

Table 3B

Interval between measles and onset of SSPE

	Interval (Yrs)	No.	%
1	< 2 yrs.	2	10.5
2	2 - 5 yrs.	5	26.3
3	6 - 8 yrs.	8	42.1
4	9 -10 yrs.	1	5.2
5	11-15 yrs.	3	15.7
Total		32	

Table 4

Presenting features

	%
Cognitive decline	93.7
Speech disturbance	59.3
Myoclonus	84.3
Gen. Tonic clonic seizures	40.6
Ataxia	46.8
Incontinence of urine	34.3
Visual impairment	18.85
Rigidity	50
Pyramidal signs	28.1

Table 5

Showing CSF IgG and Measles antibody titres

SN.	Age & Sex	CSF IgG (mg%)	CSF IgG (% of Total csf Protein)	Serum	Measles Ab CSF
1	11 M	60	100%	1:512	1:4
2	19 F	19	29.2	-	-
3	14 M	5	13.1	1:250	1:16
4	14 M	11	22.4	1:128	1:4
5	14 M	16	36.3	1:40	1:10
6	17 F	7	26.9	-	1:100
7	11 F	14	45.1	1:500	1:100
8	10 M	-	-	1:100	1:100
9	1/2M	7	100%	1:1000	1:1000
10	6 M	40.4	63.4	1:100	1:100
11	7 M	8	33.3	-	-
12	6 M	15	34	1:20	-
13	11 M	7	70	1:20	+ve
14	12 M	12	23.5	1:40	1:10
15	6 M	16	53.3	-	-
16	4 M	10	55.5	1:32	-
17	8 F	11.5	31.4	1:128	1:4
18	13 M	18.4	40.9	-	1:4
19	5 F	24	60	-	1:16

(Cont'd)

20	10 M	25	56.8	1:256	1:4
21	10 M	10	16.6	1:48	+ve
22	9 F	9	32.1	-	1:4
23	2 1/2M	5	23.8	-	1:4
24	11 F	8	21	-	-
25	7 M	-	-	-	1:4
26	9 M	24	63.1	1:16	1:1000
27	8 F	26	61.9	+	+
28	11 M	16	43.2	1:128	1:4
29	11 M	Negl.	Negl.	1:32	1:2000
30	9 M	24	50%	1:16	1:4000
31	14 M	Negl.	Negl.	1:250	1:16
32	16 M	18	32%	1:1000	1:1000

Table 6

Patients with high CSf measles antibody titres (≥ 1:1000)

Sl. No.	Age/ Sex	Titres	Age of measles	IgG %	Stage of disease	duration of disease
1	8 1/2 M	1:1000	1 yr	100	II	5 mo
2	9 M	1:1000	1 1/2yr	63.1	II	1 yr
3	11 M	1:2000	1 1/2 yr	negl	III	1 1/2 mo
4	9 M	1:4000	1 1/2 yr	50	II	1 yr
5	16 M	1:1000	N.A	32.5	II	6 mo

Table 7

Profile of patients with poor outcome

Sl. No.	Age& Sex	Measles Onset	IgG%	CSF titres	Stage	disease duration at diagnosis	Out-come
1	6 M	-	63.4	1:100	III	1 mo	Dead
2	6 M	5yr	34	-	III	5 mo	Dead
3	14 M	2 yr	36.3	1:10	II	3 mo	Bedbound
4	10 M	-	-	1:100	II	3 mo	Bedbound
5	4 M	2 yr	55.5	-	III	2 mo	Bedbound
6	10 M	8 yr	16.6	+ve	II	2 mo	Bedbound
7	9 F	-	32.21	1:4	II	6 mo	Bedbound
8	14 M	-	negl	1:16	II	9 mo	Bedbound
9	16 M	-	32	1:1000	II	6 mo	Bedbound

Table 8

Drugs used

Drug	Number
Isoprinosine	4
Levamisole	10
Valproate	12
Nitrazepam	16
Amantadine	2

Table 9

Corelation between treatment modality and outcome

S.No	CSF IgG(%)	CSF measles anitbody titre	Treatment	Outcome
1	100	1:4	VPA	Improved
2	26.9	1:100	NTZ	Improved
3	Negl	1:16	levamisole	Improved
4	63.4	1:100	NTZ + Levamisole	Dead
5	34	-	VPA + NTZ	Dead
6	36.4	1:10	NTZ	Bed bound
7	-	1:100	VPA +NTZ + Levamisole	Bedbound
8	55.5	-	NTZ + Levamisole	Bedbound
9	56.8	1:4	NTZ + Levamisole	Bed bound
10	16.6	+ve	Isoprin-osine	Bed bound
11	32.1	1:4	NTZ	Bed bound
12	32	1:1000	VPA + NTZ	Bed bound

VPA = Sodium Valproate

NTZ = Nitrazepam

DISCUSSION

The onset of SSPE usually begins in the juvenile period, the most likely ages being 5-15 yrs[6]. In our series 9.37% had onset after 15 years of age while 6.25% were below 5 yrs age (Table 1). Youngest patient was 2.5 yrs old. Males are at a greater risk than females to develop SSPE, the range of male- female ratio being 1.4 to 3.5:1[1,6]. In the present series also, this was 3:1. Males seems to have an earlier onset of the disease as compared to females (Mean age 9.37 yrs vs 11 yrs) (Table 2).

The epidemiological studies reveal that SSPE is mainly a disease of rural poor[1]. This fact is further strengthened in our study where rural: urban ratio was 7:1. The ethnic representation of SSPE was comparable to the ethnic composition prevailing in Kerala (Table 2). The relationship between measles virus and SSPE is firmly established and measles virus has been isolated from cultured brain cells of patient with this disease[17]. However in various studies positive history of measles is present in only 27% to 51.3%[20,21] cases. In our study history of measles could be obtained in 59.3% cases,

probably in the rest measles infection was subclinical or forgotten by the parent. Patient with SSPE usually have measles at an early age. 45-68% being affected below 2 years age[6]. In this series 68.4% patients had measles at or before 2 years of age (mean age 2.56 yrs). The incubation period as reported in literature is 6-8 yrs[2]. The mean incubation period in our study was 6.10, females tend to have shorter incubation period than males. (4.75 yrs vs 6.46 yrs) (Table 3). An interesting feature noted was that those who had definite history in the past had a worse prognosis than those without. In them the onset was SSPE was earlier and it progressed faster. It appears that probably those with subclinical measles infection have a milder viral load. Most of the patients presented in stage II (81.2%) with myoclonic jerks, convulsions and motor disabilities and 20 patients (62.4%) presented within 6 months of onset. In the present study the commonest presenting symptom were cognitive decline (93.7%) and myoclonic jerks (84.3%). Other common clinical features were speech disturbances, generalized seizure, ataxia and sphincteric disturbance (Table 4). Ocular signs are reported in SSPE in 50% of patients which includes chorioretinitis, cortical blindness and optic atrophy[6] while in few other studies, ocular manifestation

were not as common (<10%)[20,21]. In our study ocular manifestations were present in 18.75%. Cerebrospinal fluid (csf) examination in SSPE shows no abnormalities of pressure, cell count, total protein or sugar. Elevation of measles antibody titre and of IgG concentration in the csf is seen in all patients[2]. Prior to the measurement of csf measles antibody the presence of increased globulin was very helpful in confirming diagnosis of SSPE. In SSPE IgG concentration ranges from 10-54 mg % as compared to 5-10 mg % in normal children[6]. In the present study IgG was negligible in csf in 2 patients while in 8 patients (25%) the level was below 10 mg %, nevertheless in 7 of them csf IgG/protein ratio was significant (i.e. more than 20%). In 20, it ranged from 11-60 mg percent (Table 5).

The most consistent laboratory confirmation of the diagnosis of SSPE at present is the csf measles antibody titre. Elevation of serum measles antibody is not as specific[1]. These csf antibodies are synthesized intrathecally and not simply due to passage of serum IgG across damaged blood brain barrier[6]. CSF measles antibody titres range from 1:4 to over 1:512[6]. In our study csf measles antibody titre done in 26 patients showed significantly raised titres in 23 patients while in 3

patients titres were less than 1:4. Low csf measles antibody titres have been reported in some studies[21].

CT scan abnormalities are not detected within first year after diagnosis. Chronic patients have evidence of ventricular dilation, cortical atrophy or multifocal low density white matter lesions[6]. In our study 19 patients in whom CT scan was done, 3 patients showed lesions in the form of white matter hypodensities. Electroencephalography alterations are characteristic in SSPE, and consists of synchronous high amplitude slow wave complexes which recur at intervals of 5-15 seconds (periodic complexes). Often there is slowing of background activity. In addition to periodic abnormalities other abnormalities seen are polymorphic delta activity, intermittent frontally dominant monorhythmic slow wave activity, electrodecrement following periodic complexes, various other epileptiform discharges[18]. In the present study the periodic complexes was seen in all, 20 had slow wave complexes while other had sharp and slow wave complex. In 3 patients periodic complexes were brought out after parenteral diazepam. In 6 patients burst suppression was present. SSPE is fatal in over 95% of cases. 80% of individuals succumbing between 9 months and 3 years. Partial remission can occur at any

stage of the disease most frequently reported in stage 2 or 3 patients[1]. In our study only 37.7% (12 patients) responded to postal follow up, 2 had died, both before 9 months of onset of illness. 3 patients improved with partial deficits. One patient had visual impairment due to B/L chorioretinitis, other 2 had mild cognitive deficit. All 3 had independent existence. The improvement of disease status had no relation to csf IgG and measles antibody titre or any specific therapy (Table 8,9).

We conclude that SSPE is not uncommon in our population especially rural population. The commonest age group affected is 5-15. Males are more commonly affected. In them the disease is more severe. In about one third patients history of measles is not available, majority have measles at a median 2 yrs age and the mean incubation period is 6.10 yrs. Cognitive decline and myoclonic jerks are the hallmark of SSPE. Periodic complexes in EEG are characteristic of SSPE.

REFERENCES

1. Dyken PR : Subacute Sclerosing Panencephalitis, Current status, Neurology clinics, Vol-3, No.1, 1985: 179-195.

2. Graves MG; Subacute Sclerosing Panencephalitis, Neurology Clinics, Vol-2, No.2 1994; 267 - 280.

3. Park JE and Park K.; Measles, In textbook of Preventive and Social Medicine, 12th Ed., 1989, M/S Banarsidas Bhanot Publishers, Jabalpur; India, 118-120.

4. Saha V, Jacob John T, Mukundan P, Gnanamuthu C, Prabhakar S, Arjundas G, Sayeed ZA, Kumaresan G, Srinivas K; High incidence of subacute sclerosing panencephalitis in South India, Epidmiol. Infect, 104/1, 1990; 151-156.

5. Miller C, Farrington CP, Harbest K; The epidmiology and subacute sclerosing panencephalitis in England and Wales 1970-1989; Int. J.Epidemiol, 21/5, 1992;998-1006.

6. Peggy T. Swoveland and Kenneth P. Johnson; Subacute Sclerosing Panencephalitis and other paramyxovirus infections, In Handbook of clinical neurology, Vol. 12 (56): viral Disease, RR Mckendall ed., Elsveer science publishers B.V., 1989, 417 - 437.

7. Singhal BS, Wadia NH, Vibhakar BB and Dastur DK: Subacute sclerosing panencephalitis I, clinical aspects, Neurology India, Vol. 22, No. 2, 1974, 87 - 94.

8. Detels R, McNew J, Brody JA, Edger AH:Further epidemiological studies of subacute sclerosing panencephalitis, The Lancet, Vol 2, 1973; 11- 14.

9. Shafyi A, Lotfi J, Mirchamsy M; Subacute sclerosing Panencephalitis in Iran, Kitasato Arch - Exp.Med, 57/4, 1984; 267-271.

10. Zilber N, Rannon L et al; Measles, measles vaccination and risk of subacute sclerosing panencephalitis (SSPE), Neurology 33, 1983, 1558 - 64.

11. Halsey N.A., Modlin J.F., Jabbour JJ et al: Risk factors in subacute sclerosing panencephalitis: A case control study, Am. J. Epidemiology, 111, 1980: 415-424.

12. Risk W.S., Haddad F.S. and Chemali R; substantial spontaneous long term improvement in subacute sclerosing panencephalitis. Arch. Neurol, 35; 1978, 494 - 502.

13. Dyken PR, Nicolas S. Krawiecki, Robert H et al: The changing clinical expression of subacutre sclerosing panencephalitis in the United States., Annals of Neurology vol. No.3, 1983, 386-387.

14. Gilden DM, Rorke LB and Tanka R; Acute SSPE, Arch Neurol, 33, 1975, 644-646.

15. Dyken PR, Swift A and Du Rant RM: Long term follow up of patients with subacute sclerosing panencephalitis treated with inosiplex, Ann Neurol, 11, 1982, 359 - 364.

16. Niedermeyer E and da Silva FL ed., Electroencephalography - Basic Principles, clinical Application and Related Fields, Urban & Schwarsenberg Baltimore - Munich, (2nd Edition) 265-267.

17. J.H. Connolly, Margaret Hair et al; Measles Immunoglobulin in subacute sclerosing panencephalitis, British Med. J, 1, 1971; 23-25.

18. Markand ON and Panszi JG: The Electroencephalogram in subacute sclerosing panencephalitis, Arch Neurol, 32, 1975, 719-726.

19. Silverberg R, T. Brenner and O. Abramsky: Inosiplex in the treatment of subacutre sclerosing panencephalitis, Arch Neurol. 36, 1979, 374-375.

20. Thakare JP, Wadia RS, Deuskar NJ, Sharma VY, Kothari S, Gore MM and Ghosh SW: Subacute Sclerosing panencephalitis cases in Pune; Neurology India, 35, 1987, 333-339.

21. Khwaja GA, Gupta M, Sharma DK: Subacute Selerosing Panencephalitis; J. Ass. Phy. Ind., Vo. 39, No.12, 1991, 928-933.

22. Hall W.H. Choppin P.W: Mealses proteins in the brain tissue of patients with SSPE: Absence of M. Protein: NEJM, 304:1152-1155-1981.

IDIOPATHIC INTRACRANIAL HYPERTENSION: A RETROSPECTIVE STUDY

Idiopathic intracranial hypertension (IIH) is a syndrome characterized by increased intracranial pressure with its associated signs and symptoms in an alert and oriented patient but without any localizing neurological finding. The neurodiagnostic studies are otherwise normal except for increased cerebrospinal fluid (csf) pressure. In addition no secondary cause of raised intracranial presume is apparent. Its description dates back to 1897. Since then various terminology have been used i.e. pseudotumour cerebri, benign intracranial hypertension etc[1]. In 1980's various studies reported higher incidence of visual loss[2,3], hence the term benign was discarded and idiopathic intracranial hypertension was suggested which is more descriptive of the condition. IIH is not rare. It occurs with a frequency of 0.9-2.2 cases per 100,000 population per year[4,5,6]. It is more common in females. The annual rate in various studies is reported to be 1.6-4.3 for females of all ages and 3.3-12 for 15-44 age group per 100,000 per year. In obese women incidence further increases to 7.9-21.4 per 100,000 per year[5,6]. IIH is relatively uncommon in males. In one study males

constituted 16% of all patients and 25% of them were above 44 years of age[7].

Clinical features:

Symptoms of IIH patients are those of generalized increased intracranial pressure. The symptoms most commonly reported in IIH are headache (94%), transient visual obscuration (68%), pulsatile intracranial noises (58%), photopsia (54%) and , retrobulbar pain (44%), diplopia (38%), visual loss (30%) and retrobulbar pain on eye movement (33%) were less common accompaniments of IIH[8]. The headache is severe daily headache described as pulsatile. They are different from any previous headaches, may awaken the patient and usually last hours. Headache is usually described as worst head pain experienced. Visual obscuration are transient episodes of blurred vision that usually last less than 30 seconds and are followed by restoration of baseline visual function. The attacks are either monocular or binocular. Visual obscuration is believed to be caused by transient ischemia of anterior optic nerve or disc due to raised tissue pressure[9]. This report leads to the conclusion that transient visual obscuration are produced at the optic disc.

The signs of IIH is a result of papilloedema and resultant visual loss and sixth nerve palsy. A history of horizontal diplopia is obtained in about a third of IIH patient and 6th nerve palsy is present in 10-20%[10]. There are few case reports of patients with increased intracranial pressure of unknown cause without papilloedema[11]. In a prospective study 26% of patients at the initial visit had abnormal visual acuity[10]. There was visual loss as determined by Goldmann perimeter in 96% and by automated perimetry in 92%.

Visual loss in IIH:-

Visual loss is the only serious complication of IIH and may occur early or late. The rate of visual loss is variable, symptoms may be minimal and in many patients the visual loss may be undetected until profound. Visual impairment is most likely due to nerve conduction defect related to the stasis of axoplasmic flow found in papilloedema. Other mechanism are macular oedema, macular haemorrhage and neovascular membrane formation[19]. Nearly 50% of referral patients with IIH have been reported to have some deficit of visual function and severe deficits are reported in upto 25%[2,23]. In a recent prospective

study visual loss as determined by Goldmann perimetry occurred in 96% of 50 patients with IIH at initial evaluation[10].

Perimetry: In a prospective study of patients with IIH, visual loss in at least one eye (other than enlargement of physiological blind spot) was found in 96% of patients with Goldmann perimetry[10]. The common defects are enlargement of blind spot, loss of infero nasal position of visual field and constriction of isopters.

Risk Factor for Visual Loss:-

Corbett et al[2] found systemic hypertension to be the only statistically significant risk factor for poor visual outcome, systemic corticosteroids and raised intraocular pressure were presumed to be additional risk factors. A recent prospective study showed recent weight gain as the only factor significantly associated with worsening of vision[10].

Spinal Tap:- CSF analysis and opening pressure monitoring is mandatory for the diagnosis for IIH. Accepted csf pressure is above 200 mm of csf for non obese and above 250

mm for obese. Rarely opening pressure is normal, in such instances repeated measurements and monitoring for 30-60 minutes by an epidural transducer or via a lumbar canula may be necessary to document elevated pressure[13]. Inverse relation between csf protein a csf pressure has been reported[14]. However in a recent study low csf protein was found in only 26.% patient and there was no correlation between protein and opening pressure[15].

Pathogenesis of IIH

IIH has been associated with two fundamental defects of obscure origin, a defect in CSF absorption and increase in brain intra parenchymal water. The most popular hypothesis is that IIH is a syndrome of reduced CSF absorption[20]. Reduced conductance to CSF flow may be due to dysfunction of the absorption mechanism of the arachnoid granulation. In that case intracranial pressure must rise for csf absorption to occur. A decrease in CSF absorption in IIH has been documented by using the technique of radio isotope cisternography in several patients[28]. Increased brain water content may accompany this change. The cause of the interstitial oedema and possibly intracellular oedema is unclear but it may reduce brain compliance and prevent ventricular dilatation[19].

Vasogenic Brain Edema:- Sahs and Joynt[16] provided the first histologic evidence of extracellular edema in brain biopsy specimen. Recently increased volume of brain free water has been documented in IIH by MRI technique[17]. The increased cerebral interstitial fluid in IIH has been ascribed to transependhymal migration of CSF. Increased intracranial blood volume has been reported[18]. This phenomenon is believed to be due to obstruction of venous outflow secondary to raised pressure in subarachnoid space.

Risk Factors in IIH

Any disorder that causes decreased flow through the arachnoid granulation or obstructs the venous pathway from the granulation or to the right heart is accepted as a cause of intracranial hypertension. Arteriovenous malformation with high flow may overload venous return and elevate intracranial pressure. Steroids withdrawal and Addison's disease are clearly associated with IIH, as is hypoparathyroidism, links to other endocrine abnormalities remain unproven. Hyper-vitaminosis A is also a likely cause. Various drugs have been implicated in causing IIH- anabolic steroids, nalidixic acid, lithium, amiodarone,

sulpha antibiotics, tetracyclines, oral contraceptives etc[19]. A case control study has found strong association between IIH and obesity and weight gain during the 12 month before diagnosis[27]. A significant association of arterial hypertension and IIH and change in menstrual pattern in the previous year were noted in a case control study[27]. This study indicated wide confidence intervals for the odds ratio reflecting inadequate sample size and low statistical power for most of the variables investigated.

Diagnostic criteria for IIH

Modified Dandy's criteria[19].

1. Signs and symptoms of increased intracranial pressure.

2. Absence of localizing finding on neurological examination.

3. Absence of deformity, displacement and obstruction of the ventricular system and otherwise normal neurodiagnostic studies except for increased cerebrospinal fluid pressure (>200mm H_2O in the non obese and >250 mm H_2O in the obese patient).

4. Awake and alert patient.

5. No other cause of increased intracranial pressure present.

Neuroimaging in IIH

CT shows normal brain in IIH patients. Ventricular system may be small and cisterns may be obliterated. Moser and colleagues reported in their study using MRI that patient with IIH have a significantly higher white matter water content[21]. Weisberg[22] in his study of 28 patients reported abnormal CT scan in 36% of cases of IIH. The abnormalities included small ventricles, empty sella, enlarged cisterna magna and dilated optic nerve sheath.

Grading of Visual loss:- Various grading criteria have been developed. A simplified grading system for visual impairment in IIH by Radhakrishnan et al[1] is shown below:

Grade	Visual Symptoms	Best corrected Visual acuity	Visual Fields	
			Goldmann	Automated
Normal	None	20/20 or better	Normal field, other than enlarged blind spot	Same
Mild	May or may not be aware of defects	20/30 or better	Constriction up to 20^{o} with 14e target	No point loss >10 dB
Moderate	Aware of defects	20/40 to 20/100	Constriction>20^{o} with any target but V4e isopter at least 50^{o} only one quadrant with absolute field loss	Point loss >10 dB
Severe	Disabled by defects	Worse than 20/100	<20^{o} field with V4e target brightest target	<20^{o} field

Treatment

The therapy of IIH is dictated by the presence and progression of visual loss. Various treatment modalities are available.

1. Correction of the predisposing factors
2. Serial lumbar punctures:- If the patients symptoms resolve after the initial diagnostic lumbar puncture,

no further therapy is necessary. In those patients who continue to be symptomatic, repeated lumbar punctures have been recommended.

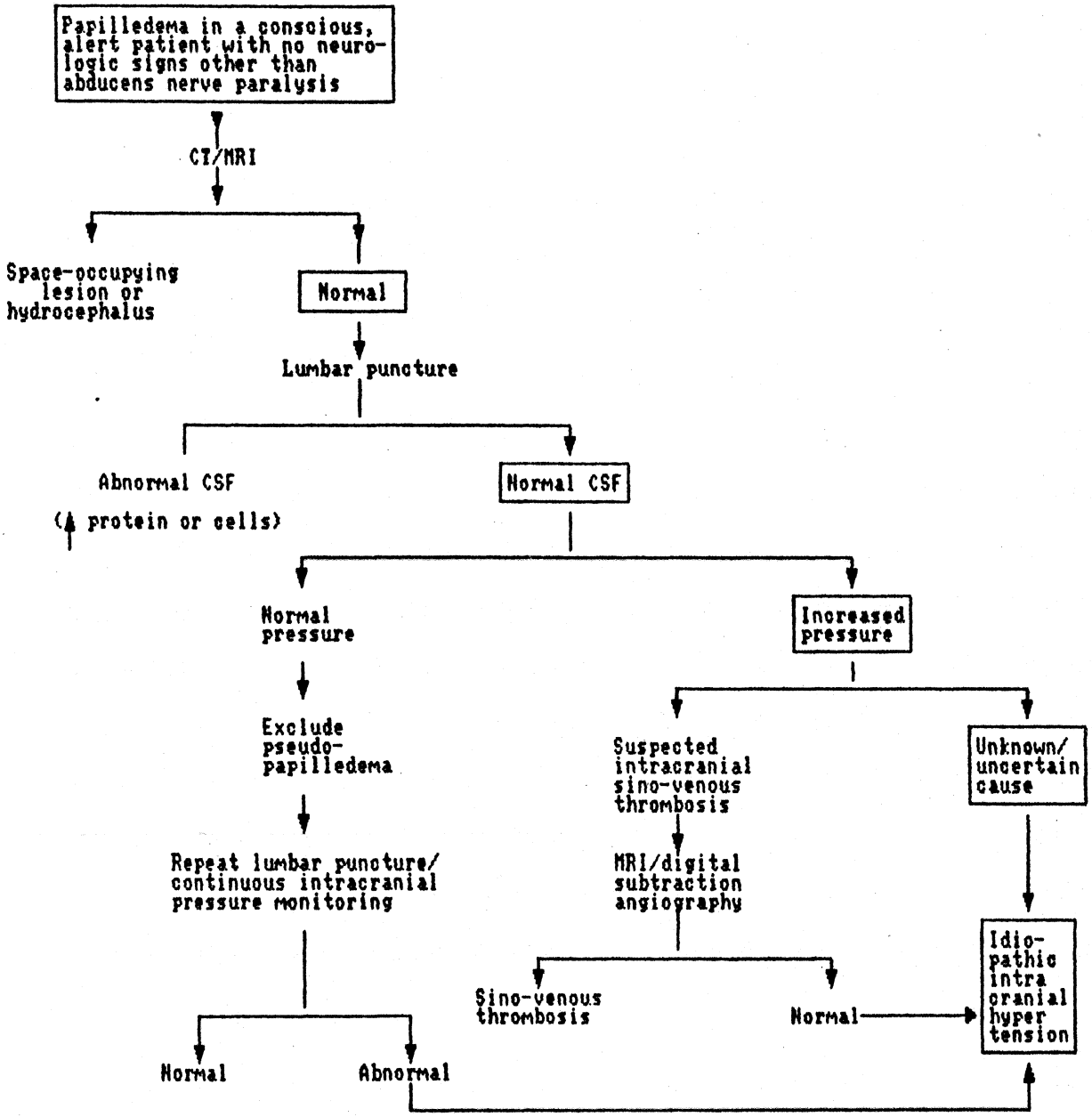
3. Acetazolamide (Diamox):- Diamox is a carbonic-anhydrase inhibitor and most frequently used. The recommended oral dose is 250 mg four times/day, increased by 250 mg/day to the maximum of 4 gm/day or till side effects develop[1].
4. Other drugs used are glycerol, furosemide.
5. Corticosteroids:- When patients are not responding to acetazolamide or develop side effects, a short course of corticosteroids 40-60 mg/day for 2-4 weeks is recommended. Prolonged use may cause recurrence or chronic course of IIH.
6. Surgery:- Surgery is indicated if visual loss progresses despite medical treatment or if initial visual loss is severe.

Surgical Options

1. Optic nerve sheath decompression or fenestration: This procedure produces immediate decompression of the optic nerve and in addition it may result in a long term filtration of CSF, surgical complication include

pupillary dysfunction, peripapillary haemorrhage, chemosis and chore retinal scaring. However, in experienced hands, the operation seems to be a safe procedure. Patients with IIH and vision loss should be offered optic nerve fenestration without delay.

2. Thecoperitoneal Shunting : Until recently thecoperitoneal shunting was generally considered the surgical treatment of choice[29]. In some reports, this procedure not only failed to halt the progressive visual loss but also carried risk of complication[30].



Algorithm for diagnosis of patients with idiopathic intracranial hypertension.

CSF = Cerebrospinal fluid
CT = Computed tomography
MRI = Magnetic resonance imaging

Aims

1. To define the frequency of occurrence of IIH in a tertiary setting.
2. To study clinical spectrum of IIH in our patients.

Material and Methods

The records of all the patients registered in Sree Chitra Tirunal Institute for Medical Sciences and Technology, Trivandrum, who were diagnosed as IIH between 1985 to 1993 were selected through computer search. Those who fulfilled modified Dandy's criteria were included in the study. Those patients who were taking cerebral decongestant at admission and had CSF pressure below 200 mm mg, otherwise fulfilling all other criteria were also included in the study. Patients with Drusen, optic neuritis,Hypertensive Retinal changes were excluded from the study.

Detailed clinical features, investigations and follow up data was studied. Visual acuity was tested by Snellen's chart and visual fields checked by Lister's perimeter. Visual impairment was graded as mild (Vision

better than 6/18), moderate (vision worse than 6/18) and severe (vision 6/60 or worse). CSF pressure was graded as severely elevated (>400 mm of csf), moderate (300-399 mm) and mild (200-299 mm of csf). Overweight (>10% of body weight) or obesity (>20% of body weight) was estimated according to prescribed norms for Indian population[26].

Results

Sixty one patients, age range 6-46 years, were seen over a period of 8 years, June 1985 to June 1993. (mean age 26.85 years and median 28 years) There were 48 females and 13 males. (F:M ratio 3.7:1). Mean age for females was 27.02 (age range 8-46 years and median 28 years) and 24.53 for males (age range 6-40 years and median 27 years). There were seven children three males and four females with a median age of 8 years. Table I shows age and sex distribution. The median duration of symptom at presentation was 1.5 months (Table-3) females presented one month earlier as compared to males.

The frequency of symptoms were headache 96.7%, diplopia 34.4%, diminution of vision 26.2%, vomiting-19.8% (Table 2). Lumbar csf presume was recorded in all. In 8 patients csf pressure was normal possibly due to cerebral decongestive treatment received prior to spinal tap. Very high csf presume (>400mm of csf) was recorded in 16.3%, moderate (300-400mm) in 22.9% and in 47.5% it was mild (200-300mm of csf). Visual impairment at initial evaluation was observed in sixteen patients. Eight had mild, 5 had

moderate and 3 had severe visual deficit. The latter were females in their third decade (Table 4). CSF pressure were 200mm, 240mm and 400 mm of csf respectively. One of them had menorrhagia. All of them recovered with medical treatment. All 5 patients with moderate visual impairment were females, 3 of them were over weight, csf pressure ranged from 200mm to 400mm of csf. One of them had empty sella on CT scan and another had hypothyroidism. All of them recovered with conservative treatment. One 33 years female was on steroid therapy at entry for suspected SLE with fluorescien angiography proven papilloedema but csf pressure was only 120mm of csf. She too had a good visual outcome. In 41 patients in whom weight record was available 21 were over weight. Table-4 shows the csf pressure, and weight in relation to visual acuity. Table 5 shows associated systemic diseases and drug ingestion in our patients. Table 6 shows the treatment modalities administered to patients. Glycerol with diamox was most frequent treatment modality. Follow up was available in 51, patients who were followed up over 1 month to 18 months. Within one month from diagnosis and start of treatment 39.2% patients were asymptomatic but papilloedema resolved in only 17.6%. By six months 88.2% were asymptomatic and papilloedema had resolved in 47%.

Papilloedema resolved in 36 patients (70.5%) with a modal resolution time of one month. Papilloedema persisted in 13 (25.4%) while 2 had bilateral optic atrophy. Table 8 shows follow up in our patients.

Table 1: Age and Sex distribution in our patients

Age	Sex		Total
	M	F	
0-9	3	1	4
10-14	0	4	4
15-19	0	6	6
20-24	0	9	9
25-29	6	5	11
30-34	2	13	15
35-39	1	5	6
40-44	1	2	3
45-49	0	3	3
		Total = 61	

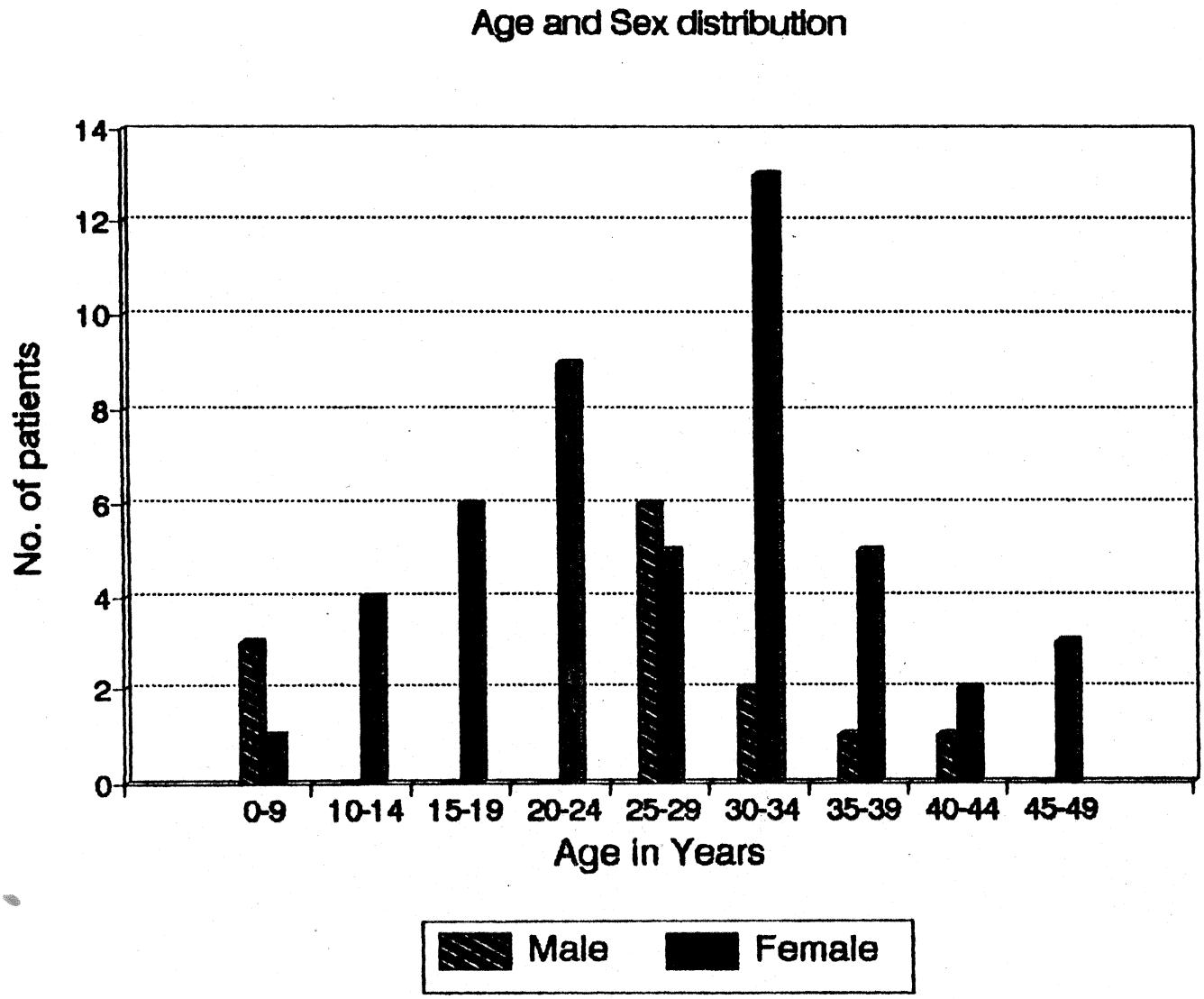

Age and Sex distribution
No. of patients
14
12
10
8
6
4
2
0
0-9
10-14
15-19
20-24
25-29
30-34
35-39
40-44
45-49
Age in Years
Male
Female

Table 2: Presenting symptoms

		Percentage
Headache	-	96.7
Diplopia	-	34.4
Diminision of vision	-	26.3
Visual Obscurations	-	16.3
Vomiting	-	22.9
Tinnitus	-	8.2
Retro orbital pain	-	3.2
Deafness	-	3.2
Photophobia	-	3.2

Table 3: Duration of symptoms at presentation

Duration (month)	No M	No F	Total	%
0.5	0	6	6	9.8
1	4	19	23	37.7
1.5	1	1	2	3.2
2	2	6	8	13.1
3	2	2	4	6.5
4	0	2	2	3.2
5.5	1	0	1	1.6
6	0	4	4	6.5
7	2	1	3	4.9
>12	1	7	8	13.1

Median - 1.5 Total = 61 Mode - 1 yr.

Table - 4: Weight and CSF Pressure in relation to visual impairment

	S. No.	Age & Sex		Wt	csf press (mm of csf)
	1.	12	F	N	200
	2.	30	F	obese	260
Mild	3.	25	M	-	280
Visual	4.	46	F	obese	260
Imp.	5.	13	F	-	160
	6.	44	F	-	280
	7.	30	F	under weight	150
	8.	6	M	N	110
	9.	40	M	obese	350
moderate	10.	20	F	-	200
visual	11.	31	F	obese	400
Imp.	12.	32	F	obese	400
	13	33	F	-	120
	14	20	F	-	200
Severe					
Visual	15	29	F	over weight	240
Imp.	16	20	F	-	240

Table - 5: Associated conditions in our patients (N - 61)

	No.
Hypothyroidism	2
Vascular headache	7
Hypertension	4
Otitis media	4
Epilepsy	1
Aortic regurgitation	1
Asthma	2
SLE	2
Pregnancy	1
Drugs	6
Vit A.	2
Corticosteroids	2
Oral Contraceptive Pill	2

Table - 6: Treatment modes in our patients

Treatment	No	%
G+D	30	49.1
Oral G alone	8	13.1
D alone	9	14.7
G + D+ P	5	8.1
P alone	3	4.9
G + L	2	3.2
G + P	3	4.9
CSF drainage	1	1.6
	Total 61	

G - Glycerol

D - Diamox

P - Prednisolone

L - Lasix

Table-7: Multivariate chart of visual impairment with associated variables with follow up)

S. N.	Age & Sex	VA R	VA L	csf pr.	wei-ght	Follow up vision Improve(mo)		Papillad Improve(mo)
1.	32 F	6/9	6/24	400	OW	1		7
2.	12 F	6/6	6/9	200	N	2		
3.	30 F	6/6	6/9	260	OW	2		2(optic atrophy
4.	25 M	6/9	6/6	280	-	-	No.F.up	-
5.	20 F	6/36	2/60	240	-	-	No.F.up	-
6.	29 F	2/60	6/60	400	N	8		8
7.	46 F	6/9	6/6	260	OW	-	No.F.up	-
8.	13 F	6/9	6/9	160	-	-	No.F.up	-
9.	40 F	6/18	6/9	350	OW	1		1
10.	44 F	6/12	6/12	280	-	-	No.F.up	-
11.	30 F	6/9	6/9	150	OW	3		9
12.	20 F	6/9	6/9	200	-	1.5		4
13.	31 F	6/18	6/9	400	OW	1.5 mo		Pap.persisted upto 1 1/2 yrs
14.	6 M	6/6	6/9	110	N	1		1
15.	20 F	6/60	6/60	200	-	4		4
16.	33 F	6/12	6/18	120	-	10		Pap.persisted upto 1 1/2 yrs

Table 8: Follow up (N-51)

F.up duration (month)	No Symptom	%	No Papilloedema	%
1	20	39.2	9	17.6
2	8	15.6	3	5.8
3	8	15.6	6	11.7
4	3	5.8	3	5.8
5	4	7.8	3	5.8
6	2	3.9	-	
7	1	1.9	3	5.8
8	1	1.9	2	3.9
9	-	-	1	1.9
>12	4	7.8	6	11.7
	Total 51		Total 36	

Papilloedema persisted - 13

Optic atrophy - 2

Total - 51

DISCUSSION

Sixty one patients with IIH were treated between 1985 to 1993 (8 yrs) i.e. 7.6 per year. The incidence of IIH is reported to be 0.9-2.2 per 100,000 per year[4,5,6]. For Kerala state with a population of 30 million, the number of cases annually works out to be 27-66. Being a tertiary referral centre, we see only a small fraction of these cases. The mean age was 26.85 yrs which is concordant with study by Bulens et al[25], The commonest age group affected was 25-34 (Table I) which correlated well with previous studies[24,25]. IIH is more common in females especially obese. In the present series male, female ratio was 1:3.7. Males comprised 21.3% of total which is close to the incidence reported by Digre et al[7]. In a population based study, Radhakrishnan et al[5] ascertained only 5 males out of 81 patients (6.1%).

Duration of symptoms prior to diagnosis in different studies varies from few days upto 18 months[5,25]. In the present study nearly half of the patients (Table-3) presented within 1 month or before. 13.1% had symptoms for more than 12 months. The commonest symptoms were headache (96.7%), diplopia (34.4%) and visual impairment (26.2%).

Frequency of visual impairment correlates well with others studies[5,8,10,25] however in the present study frequency of visual obscuration was much less (16.3%). The prognosis for vision in most patients with I.I.H is excellent however, visual loss may occur. Corbett et al[2] followed up 57 patients of I.I.H for 5 to 41 years, severe visual loss in one or both eyes occurred in 14 patients. In another prospective study[10] 2 out of 50 (4%) became blind in both eyes. In the present study 3 out of 16 had severe visual impairment at onset while 5 had moderate visual impairment. Severity of visual impairment did not correlate with csf pressure. None of the 11 patients in whom follow up was available had residual visual deficit. In 8 of them vision normalized within 4 months. 2 patients developed optic atrophy during follow up, but suffered no visual impairment. Such a good visual outcome in our patients may be due to early treatment and paucity of risk factors.

Very high CSF pressure was recorded in 16.3% (>400mm of csf). In 8 (13.1%) csf pressure recording was normal, all were taking cerebral decongestant at entry, in 2 of them flourescien angiography confirmed papilloedema. In one there was evidence of raised intracranial pressure in X-ray skull while in the rest there was unequivocal evidence of papilloedema when examined by neurologist and

ophthalmologist.Rarely opening pressure in IIH may be normal. In such instances prolonged csf presume monitoring for 30-60 minutes is recommended[13]. Obesity especially recent gain in weight is an important association and risk factor for IIH[19]. Nearly half of the patients in whom weight was recorded were overweight. Much higher frequency of obesity (71-94%) have been reported in various other studies[5,10]. Even though men with IIH are less likely to be obese than women, they tend to be more obese than control subjects[7]. In the present study weight was available in 3 of 13 males, all were overweight. There was no correlation between weight, csf pressure and visual impairment in the present study. The commonest associated medical conditions noted were vascular headache (11.4%), hypertension (6.5%) and otitis media. (6.5%).

Treatment and follow up

All but two responded to conservative treatment. Combination of glycerol and diamox was most common treatment modality. Steroids alone or in combination with other drugs were used in only 15%. This further emphasizes that most cases can be managed successfully without steroids or surgical intervention. One patient a 28 yrs old male required csf drainage by multiple lumbar punctures.

His csf pressure was 360 mm. Another was a 40 yrs obese female with csf pressure of 350mm who required thecoperitoneal shunt. Both had uneventful recovery. Follow up was available for 51 patients (83.6%). Patients were followed up for 1 month to 18 months. Symptomatic improvement was seen within one month in 39.2% most responded within 6 months (88.2%). More objective evidence of response in the form of resolution of papilloedema was present in 36 out of 51 patients (70.5%), while it persisted throughout follow up in 13 (25.4%). 2 developed optic atrophy. By one month resolution of papilloedema was seen in only 17.6% and in 47% within 6 months. indicating that regression of symptoms occurs earlier to resolution of papilloedema. This fact must be taken into consideration while treating the patients in follow up. We conclude that frequency of IIH seen is much less as compared to its incidence in population based studies. Females in age group 20- 40 are predominantly affected, most patients are referred early on our set up. Although visual impairment is not uncommon, visual outcome is uniformly good, CSF pressure does not correlate with visual impairment. Symptomatic improvement occurs earlier than resolution of papilloedema. This should help in modifying drug treatment in follow up.

REFERENCE

1. Radhakrishnan K, Ahlskog JE, Garrity JA, Kurland LT; Idiopathic Intracranial Hypertention; Mayo clinic Proc. 69: 169-180, 1994.

2. Corbett JJ, Savino PJ, Thompson HS, Kansu T, Schatz NJ, Orr-LS, Hopson D: visual loss in pseudotumor cerebri. Follow up of 57' patients from 5 to 41 years and profile of 14 patients with permanent severe visual loss. Arch Neurol,39:461-74. 1982.

3. Orcutt JC, Page NG, Sanders MD. Factors affecting visual loss in benign intracranial hypertension. Ophthalmology 91, 1303-1312, 1984.

4. Durcan FJ, Corbett JJ, Wall M. The incidence pseudotumor cerebri: Population studies in Iowa and Louisana. Arch Neurol 45, 875-877, 1988.

5. Radhakrishnan K, Thacker AK, Bohlaga NM, Maloo JC, Gerryo SE. Epidemiology of idiopathic intracranial hypertension: a prospective and case-control study. Journal of Neurol Sci 116: 18-28, 1993.

6. Radhakrishnan K, Ahlskog JE, Cross SA, Kurland LT, O'Fallon WM. Idiopathic intracranial hypertension (Pseudotumor cerebri): descriptive epidemiology in Rochester, Minn, 1976 to 1990, Arch Neurol. 50, 78-80,1993.

7. Digre K.B, Corbett JJ: Pseudotumor cerebri in men, Arch Neurol 45, 866-872, 1988.

8. Wall M, Giuseffi V, Rojas PB: Symptoms and Disease association in pseudotumor cerebri: A case control study; Neurology 39 (suppl): 210, 1989.

9. Sadun A, Currie J, Lessell S: Transient visual obscurations with elevated optic discs. Ann Neurol 16: 489-494,1984.

10. Wall M, George D: Idiopathic intracranial hypertension. A prospective study of 50 patients; Brain, 114: 155-180, 1991.

11. Lipton HL, Michelson PE: Pseudotumor syndrome without papilloedema; JAMA 220: 1591-1592, 1972.

12. Verplanck M, Kaufman DI, Parsons T, Yedavally S, Kokinakis D: Electrophysiology versus psychophysics in the detection of visual loss in pseudotumor cerebri; Neurology 38 (11) 1789-92, 1988.

13. Gucer G, Viernstein L: Long term intracranial pressure recording in the management of pseudotumor cerebri, J. Neurosurg, 49, 256-263, 1978.

14. Chandra V, Bellur SN, Anderson RJ: Low CSF protein concentration in Idiopathic pseudotumor cerebri, Ann. Neurol 19: 80-82, 1986.

15. Johnston PK, Corbett JJ, Maxner CE. Cerebrospinal fluid protein and opening pressure in idiopathic intracranial hypertension(Pseudotumor cerebri); Neurology 41: 1040-1042, 1991.

16. Sahs AL, Joynt RJ: Brain swelling of unknown cause; Neurology 6: 791-803, 1956.

17. Sorensen PS, Thomas C, Gjerris F, Henriksen O. Brain water accumulation in pseudotumor cerebri demonstrated by MR imaging of brain water self diffusion; Acta Neurochir suppl (Wein) 51: 363-365, 1990.

18. Mathew NT, Meyer JS, Ott EO. Increased cerebral blood volume in benign intracranial hypertension; Neurology 25: 645-649, 1975.

19. Wall M: Idiopathic Intracranial Hypertension. In Neurologic clinics vol 9, No.1, 73-95, 1991.

20. Fishman RA: Pathophysiology of Pseudotumor cerebri An unsolved puzzle, Arch Neurol 41, 257-258, 1984.

21. Moser FG, Hilal S K, Abraham G; MR Imaging in pseudotumor cerebri, Am. J. Radiol 150, 903-909, 1988.

22. Weisberg LA: Computed tomography in benign intracranial hypertension; Neurology 35, 1075-1078, 1985.

23. Sorensen PS, Krogasaa B, Gjerris F: Clinical course and prognosis of pseudotumor cerebri: a prospective study of 24 patients; Acta Neural Scand 77, 164-172, 1988.

24. Johnston I and Peterson A: Benign Intracranial hypertension-I; Diagnosis and prognosis; Brain 97, 289-300, 1974.

25. Bulens C, De Vries WAEJ, Van Crevel H : Benign Intracranial Hypertension; A retrospective and follow up study; J. Neurol sci, 40: 147-157, 1979.

26. Gopalan C, Combating under nutrition. Basic issues and practical approaches, (Nutritional foundations of India), Special publication series-3, 292, 1980.

27. Ireland B, Corbett JJ, Wallace RB : The search for causes of Indiopathic Intracranial Hypertension A preliminary case control study Arch-Neurol: 47: 315-20,1990.

28. Gjerris F, Sorensen PS, Vorstrup S, Paulson OB, Intracranial pressure, conductance to cerebrospinal fluid outflow, and cerebral blood flow in patients with benign intracranial hypertension (Psendotumor cerebri) Ann. Neurol: 17:158-162,1985.

29. Johnston I, Besser M, Morgan MK, Cerebrospinal fluid diversion in the treatment of benign intracranial hypertension J Neurosurg. 69:195-202, 1988.

30. Rosenberg M, Smith C, Beck R, Cerbett J, Sergott R, Savino P et al. The efficacy of shunting procedures in psendotumor cerebri (abstract). Neurology 39:(suppl 1), 209, 1989.