Guillain Barre Syndrome clinical profile & the efficacy of different modalities of treatment

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Summary :

Seventy five patients with Guillain-Barre syndrome (GBS) admitted to 3 centers in Baghdad between January 2000 to June 2002 at ages 6months-15 years were studied for various clinical and cerebrospinal fluid aspect , complications ,outcome of illness in different modalities of treatment (plasma pheresis, steroid therapy , supportive care) .
It was found that 44 of them were males and 31 were females in ratio male:female 1.4:1 .
It was found that 60% of them had upper and lower limbs paralysis and 40% had only lower limbs paralysis .76% had sensory changes , 33% had cranial nerves involvement , 20% had meningeal irritation , 68% had elevated protein in cerebrospinal fluid.
Respiratory involvement was found in 28 patients (37%) , 21 patients needed ventilatory support .Mortality rate was 4% (3 patients) , all died in respiratory care unit (RCU) due to cardiac complications and secondary infections .
Sixty percent of the patients had upper respiratory tract infection preceded the illness , 20% of patients had gastrointestinal problems , 2.67% of patients had history of DPT and oral polio vaccine , 10.67% of the patients had fever , 6.67% of the patients had no any previous pathology.
Autonomic disturbances was found in 42.6% of the patients .
Fifteen patients received plasma pheresis(recovery rate 93%) .
Twenty patients received steroids (recovery rate 75% ,mortality rate 10%), 40 patients received supportive (recovery rate 88%,mortality rate 2%) .

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Introduction

Guillain-Barre syndrome (GBS), an acute flaccid paralysi or ascending quadriparesis in an infant or child constitute a very important pediatric neurological emergency .The GBS is the most frequent cause of flaccid paralysis.It is primarily an autoimmune postinfectious demyelinating peripheral nervous system process [1]. Although GBS previously viewed as unitary disorder with variation, it currently viewed as group of syndromes with several distinctive subtypes like acute inflammatory demyelinating polyradiculoneuropathy, acute motor axonal neuropathy, acute motor sensory axonal neuropathy, Miller-Fisher syndrome and perhaps others[2] . Because of potential for acute respiratory compromise, any child suspected to having GBS need immediate hospitalization [3] . Although most children with GBS have relatively benign clinical course, some become very ill and required intubation with intensive care monitoring [1].

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PATIENTS AND METHODS :

A prospective study of 75 patients was conducted , referred patient from different medical centers in Iraq examined and follow up done for them in 3 center (Al Mansour teaching hospital : 40 patients , RCU in Al shaheed Adnan hospital :22 patients , national bank of transfusion center :13 patients) in a period from January 2000 to June 2002 . The variables for analysis in the study include child factors (age , sex , date of admission , date of discharge , clinical features which include { antecedent infections , motor nerve weakness , sensory , autonomic , cranial nerve involvement} , respiratory muscle involvement , RCU admission , asymmetry of weakness , meningeal irritation , CSF protein , mortality rate , full recovery or mild neurological impairment) as shown in table I and III . Lab studies were done to all patients including: C.S.F. study except in ten patients were 7 of them the family refuse LP and in 3 patients the LP was traumatic , the sample of CSF tested for(sugar ,protein and cells) , electrocardiography , blood tests (complete blood count , erythrocytes sedimentation rate , blood sugar , blood urea and electrolytes) and general urine examination . Neurophysiological studies were performed in 20 patients that recording nerve conduction velocity (NCV) and electromyography (EMG) .Other

Discussion:

Clinical, laboratory and Neurophysiological aspect with different modality of treatment in 75 child admitted to 3 centers in Baghdad was adequately evaluated in a period from January 2000 to June 2002. The age and sex of the patients were analyzed that showed ; 44 patients were male (58.67%) and 31patients were female (41.34%) in a ratio of male :female (1.4:1) as shown in table I , that agree with other studies which show the male predominant like that which done by Saleh F- Al Ajlouni who found that male :female ratio 1.6:1 , and other study done by Tabaraki which show the same result , but the ratio is differ in study done by Zielinska who found female predominant (1.13:1) [5,] .Neurological manifestations that preceding medical illness (as shown in table II) , the upper respiratory tract infections in 45 patients (60%) , gastrointestinal tract infection in 15 patients (20%) , DPT and oral polio in 2 patients (2.67%) , fever only in 8 patients (10.67%) and in the last 5 patients (6.67%) no preceding events were found .this agree with other study which show upper respiratory tract infection in the majority of the cases , then gastrointestinal tract infections [6,] while Zhonghua suggest that occurrence of GBS may correlate to the infection with compylobacter jejuni and poor personal hygiene in children [8] . All patients in this study show motor weakness , in 71 patients (95%) the weakness was symmetrical ascending but it is asymmetrical in 4 patients (5%) , both upper and lower involvement was noted in 45 patients (60%) but weakness in lower limb only was found in 30 patients (40%) , this results was similar to the results of study done by Asbury AK and Cornblath which show that all patients had flaccid symmetrical paralysis[7,9] . Sensory changes seen in 67 patients (89%) , 25 patients show cranial nerves involvement (33%) .this results is approximately similar to other study done by Saleh F Al-Ajlouni and M.N Al-Nidawi which show cranial nerves involvement in 38.4% and 37.1% respectively [7,10] .The possibility of Miller- Fisher syndrome was raised in 3 patients that presented with acute ataxia and areflexia but without ophthalmoplagia and reviewing electrophysiological features which show demyelinating sensory motor polyneuropathy which met criteria of ataxic GBS while this possibility of Miller-Fisher syndrome was raised only in one patient in a study done by M.N Al Nidawi in 1994 [10] . Respiratory involvement was found in 28 patients (37%) and 21 patients of them need ventilatory support and other 7 patients need close observation only, this was similar to study done by Winner JB and also similar to the study which is done by Saleh F Al Ajlouni [7,10,11,12] . Meningeal irritation and neck stiffness were found in 15 patients (20%) while in study done by Kaal ,-EG who found that the classical symptoms such as flaccid paralysis and areflexia are not always predominant instead pain is often the most prominent symptoms along with meningism [10] . The mortality rate was 4% , all (3 patients) died in the respiratory care unit due to the complications of illness or secondary bacterial infections , this rate was approximately less than that which was found in a study done by Tabaraki (7%) [4] . Autonomic involvement was found in 32 patients (42.6%) in form of techycardia (in 20 patients , 27%) , hypotension (10 patients ,13%) and urine retention (in 2 patients , 2.6%) and this agree with the results of the study which is done by Asahina M in Japan . The patients were distributed allover the months with 2 peaks (in winter and summer) , this agree with a study done by Paradiso –G that show the high risk is mainly in summer and winter [15] . In the treatment of our patients , those who presented early in the first week who treated by plasma pheresis get rapid recovery and early mobilization but the patients who presented later in the illness who also treated by plasma pheresis show poor response , this agree with study done by Bruck -I who found that faster clinical improvement at day 5 with plasma pheresis treated group but after 20 days in those who treated by supportive measures only [16] while Zielinska-M report that the influence of the treatment method on clinical course of the disease and he found that the plasma pheresis applied to patients with severe GBS significantly reduce the period of the paralysis and enabling early rehabilitation and faster mobilization of the patients while the application of steroids did not have significant influence on the course and final effect of treatment of GBS [5] while in other study which done by Graf-WD who concluded that no much difference between Ig , plasma pheresis or supportive measures if used in late phase in very severe illness but effective only (especially plasma pheresis or Ig) when given to certain patients very early in the course of the illness [17] . Those patients who were treated by steroid , most of them presented with rapidly progressive severe illness , so steroids tried in their treatment because the plasma pheresis was not available but their outcome in comparison to the supportive care was no much differ , instead of that some of them show prolong course of illness , this is similar to some studies like that which is done by Kenneth [18,19] and other one done by Walton whom their conclusion was that there is no beneficial effect was gained from giving steroid in comparison to those who were not given. Table VII show Progression of the illness according to the modality of treatment in which show the efficacy of plasmapherisis in management of G.B.S.

Appendix 1 (9)
Diagnostic criteria of Guillain-Barre syndrome after Asbury and Cornblath

1. Features required for diagnosis :
a-Progressive motor weakness of more than 1 limb.
b-Loss of tendon jerks .
2- Features strongly support the diagnosis :
a-Clinical features :
1-progression over 4 weeks .
2-relative symptoms of weakness .
3-mild sensory signs or symptoms .
4-cranial nerves involvement .
5-recovery usually beginning 2-4weeks after progression stop.
6-autonomic dysfunction .
7-absence of fever at the onset of neurological symptoms .
b-CSF features :
1-CSF protein raised after the first week of symptoms .
2-counts of 10 or fewer mononuclear leukocytes x 106 /L .
c-Electrodiagnostic features :
Reduction of conducting velocity ,conduction block or abnormal temporal dispersion ,increased distance latency or abnormal F wave in more than one nerve.
3- Features casting doubt on the diagnosis :
a-Marked persistent asymmetry of weakness .
b-Persistent bladder or bowel dysfunction .
c-Bladder or bowel dysfunction at onset .
d-Presence of polymorphonuclear leukocyte in the CSF .
e-Sharp sensory level .
4-Features that rule out the diagnosis :
a-Indication of any metabolic ,infectious or disease associated with polyneuropathy .
b- Occurrence of a purely sensory syndrome .

Appendix 2 (9)
0- No movement .
1- Flicker or trace contraction .
2- Active movement with gravity eliminated .
3- Active movement against gravity .
4- Active movement against gravity and resistance .
5- Normal power .

Appendix 3 (20)

Plasmapheresis as first – line therapy

- Cryoglobulinemia
- Auti - GBM disease
- GBS
- Hyperviscosity syndrome

Microangiopathie thrombocytopenia (TTP/HUS)
Homozygous familial hypercholesterolemia
Myasthenia gravis crisis
Overdose with certain drugs
Coagulation factor inhibitors
Plasmapheresis as adjunctive treatment
Rapidly progressive glomerulonephritis
Systemic vasculitis
Multipl myeloma with renal involvement
SLE

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Table (1) Shows The Prevalence Of Enuresis In Each Age Group.

Age	Enuretic		Non - enuretic		Total
	No.	%	No.	%.	
5-7	65	24.4	205	75.6	270
8-10	30	25	115	75	145
11-13	15	18	70	72	85
5-13 all	110	22	390	78	500

Table (2) Shows The Prevalence Of Enuresis In Each Age Group In Relation To Sex (Male, Female) Ratio

Age group	male					female				
	Total	Non-enuresis		enuresis		Total	Non-enuretic		enuresis	
		No.	%	No	%		No.	%	No.	%
5-7	165	120	76	40	24	118	92	76	26	21
8-11	65	49	76	16	24	64	48	66	16	25
11-13	50	46	92	4	8	38	30	86	8	21
5-13 all	280	220	76	60	21	220	170	84	50	21