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Anticipating Mechanical Ventilation In Children With Guillain –Barre Syndrome And Improving Outcome Of The Illness

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Summary:

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Background: *Acute inflammatory demylinating polyneuropathy (Guillain-Barr'e syndrome) is the commonest cause of acute neuromuscular paralysis in most countries. The onset of symptoms in patients with GBS can either be acute or sub acute .Ventilatory failure is primarily caused by inspiratory muscles weakness although weakness of abdominal and accessory muscles of respiration, retained airway secretion leading to aspiration and atelectasis are all contributing factors. Nutritional support of critically sick children is important for metabolic maintenance and tissue repair.*

Methods: *To identify clinical and respiratory features associated with progression to respiratory failure in children with GBS. Twenty two consecutive children with severe (Rapidly progressive) GBS admitted to the RICU in Children Welfare Teaching Hospital-Medical City –Baghdad, between July 2004-July 2006, were studied .Other 22 children with sub acute GBS who didn't receive mechanical ventilation admitted in the same period to the same hospital as a control group.*

Results: *There is significant association between each of bulbar, autonomic neuropathies and low SiO2 at admission with severe GBS .There is significant reduction in duration of mechanical ventilation and duration of RICU stay achieved by the use of Freamine amninoacid solution as partial parenteral nutrition.*

Conclusion: *Progression to mechanical ventilation was highly likely to occur in those patients with rapid disease progress (less than one week) bulbar dysfunction and dysautonomia, and low SiO2 at admission. Partial parenteral nutrition may reduce the duration of mechanical ventilation and RICU stay by about 3 days.*

Keywords: *GBS; mechanical ventilation; children.*

Introduction:

Acute inflammatory demylinating polyneuropathy (Guillain-Barr'e syndrome) is the commonest cause of acute neuromuscular paralysis in most countries. Incidence figures vary from 0.4-4/100000 population/year[1].Approximately one third of patients with Guillain-Barr'e syndrome develop respiratory failure requiring mechanical ventilation .Ventilatory failure is primarily caused by inspiratory muscles weakness although weakness of abdominal and accessory muscles of respiration ,retained airway secretion leading to aspiration and atelectasis are all contributing factors [2] .The combination of multiple clinical factors culminates in neuromuscular and respiratory failure in patients with Guillain-Barr'e syndrome [3]

Treatment of Guillain-Barr'e syndrome is mainly symptomatic and supportive. Regular measurement of respiratory function and consideration of elective ventilation are vital, bulbar dysfunction may require nasogastric feeding, chest and limb physiotherapy are helpful. Specific treatment is now by the use of intravenous immunoglobulin (IVIG)or plasmapharesis .Studies have shown to be equally effective .The former is considerably easier to administer[4,5] .Rapidly progressive ascending paralysis is treated with IVIG for 2,3,or 5 days ; plasmapharesis ,steroid ;and immunosuppressive drugs are alternatives if IVIG is ineffective [6] . Nutritional support of critically sick children is important for metabolic maintenance and tissue repair .Parenteral nutrition is indicated when oral or enteral nutrition is impossible, insufficient or contraindicated. In areas where facilities for total parenteral nutrition are not available, partial parenteral nutritional can be made and can provide a satisfactory support for up to 2 weeks [7].

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Materials And Methods:

To identify clinical and respiratory features associated with progression to respiratory failure in children with Guillain-Barr'e syndrome. Twenty two consecutive children with severe (Rapidly progressive) Guillain-Barr'e syndrome admitted to the Respiratory Intensive Care Unit (RICU) in Children Welfare Teaching Hospital(CWTH)-Medical City –Baghdad, between July 2004-July 2006 ,were studied .They fulfilled the Asbury and Cornblath clinical criteria for the disease which include progressive motor weakness of more than one limb, and loss of tendon jerks and confirmatory features of CSF protein raised after the first week of illness and count of 10 or fewer mononuclear leucocytes X10 ^{6}/L [8] .Variables taken included :Age, sex, antecedent event ,duration of

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weakness before admission ,motor, sensory ,autonomic and bulbar involvement ,SiO2 at admission ,CSF protein and CSF cells.

Clinical and respiratory features of 22 children receiving mechanical ventilation compared with other 22 children with sub acute Guillain-Barr'e syndrome who didn't receive mechanical ventilation admitted in the same period to the same hospital as a control group. Supportive management including physiotherapy and nutritional therapy with partial parenteral nutrition as facilities for total parenteral nutrition was not available and the enteral nutrition was not satisfactory, an amino acid solution was administered through a peripheral vein for 7-10 days duration. The amino acid solution used was (Freamine Baxter ATC: B05BA01) which is a pediatric amino acid solution designed to achieve amino acid levels seen in the cord blood premature or full term infant, a well balanced and complete amino acid mixture that contain a quite high amount of taurine, cysteine ornithine and high strength of amino acid content 8.5 % with out electrolyte [7,9].Duration of ventilation and duration of stay in the RICU were studied between the group who received Freamine and the second group who did not receive it as the second control group. Statistical analysis was done by using SPSS version 11 program and Chi-square test was used and a P. value < 0.05 was considered significant.

Results:

The sex distribution of the patients with severe Guillain-Barr'e syndrome showed 14 males (63.6%) and 8(36.4%) females with a male/ female ratio = 1.75/1. The mean age was 51.45 ± 43.73 months with an age range between 5 months and 108 months. The mean duration of weakness before admission was 4.14 ±1.73 days with a rang of 1 to 6 days. Antecedent event was recorded in 17 patients (77.3%), 15 cases with history of upper respiratory tract infection (URTI), one with gastrointestinal tract infection (GITI) and one had history of DPT vaccine administration 2 weeks before the start of weakness. All patients had ascending motor paralysis, 8 (36.4%) had sensory impairment, and 10 (45.5%) showed manifestations of autonomic nervous system involvement, (3 with hypertension, 4 with tachycardia, 3 with GIT in the form of constipation, 3 with urinary retention and one with breadycardia), taking in consideration that some of them developed more than one autonomic manifestation like the coexistence of hypertension and tachycardia. As regard cranial nerves (bulbar) involvement it was recorded in 17 (77.3%).The mean oxygen saturation at admission was 87.18 ±5.64. The mean CSF protein was 88.43± 66.42 mg/dl and the mean CSF cells were 5.2±4.5 x10 ^{9}/l. All were mechanically ventilated. Eighteen patients were treated with immunoglobulin (IVIG) and corticosteroid and the remaining 4 patients did not receive corticosteroid and were treated with IVIG alone.

Fourteen patients with severe Guillain-Barr'e syndrome received partial parenteral nutrition, their mean duration of mechanical ventilation was 12.5 ±53 days and their mean duration of stay in RICU was 25.85 ± 6.03 days, while 8 patients did not receive partial parenteral nutrition, their mean duration of mechanical ventilation was 16.14 ± 483 days and their mean duration of stay in RICU was 29.25 ±7.3 days . There is significant difference between the 2 groups regarding mean duration of mechanical ventilation (P.value =0.014) and mean duration of stay in RICU (P.value=0.03).All children including those with severe and subacute Guillain-Barr'e syndrome were discharged with improvement and no death was recorded.

Table-1: The associations of risk factors (frequencies) with severe Guillain-Barr'e syndrome

Factors	Severe GBS with mechanical ventilation		Subacute GBS without mechanical ventilation		P.Value
	No	%	No	%	
Male sex	14	63.3	12	54.5	0.5
Antecedent events	17	77.3	12	54.5	0.05
Sensory neuropathy	8	36.4	5	22.7	0.3
Autonomic neuropathy	10	45.5	6	27.3	0.03
Bulbar neuropathy	17	77.3	8	36.4	0.003

Table-2: The associations of risk factors (means) with severe Guillain-Barr'e syndrome

Factors	Severe GBS with mechanical ventilation		Subacute GBS without mechanical ventilation		P.Value
	Mean	SD	Mean	SD	
Age (months)	51.54	34.73	63.00	35.1	0.1
%O2 Saturation at admission	87.18	5.64	96.3	2.71	0.03
CSF Protein (mg/dl)	88.43	66.47	88.47	69.o	0.5

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Table-3: The effect of Freamine supportive treatment on the course of severe Guillain-Barr'e syndrome

Duration	Freamine treated group		Non Freamine treated group		P. Value
	Mean	SD	Mean	SD	
Duration of artificial ventilation(days)	12.5	5.37	16.14	4.80	0.01
Duration of RCU stay (days)	25.85	6.03	29.25	7.30	0.03

Discussion:

The onset of symptoms in patients with Guillain-Barr'e syndrome can either be acute or subacute .In a large multicentre study the mean time to reach nadir is 12 days [5], and all patients in the present study had needed mechanical ventilation in less than one week from the onset of illness, hence they were considered as having acute severe (rapidly progressive) Guillain-Barr'e syndrome.

Males appeared to be affected more commonly (males/females =1.7:1). This was noted in other studies [5, 10], but male sex shows no significant association with the severity of Guillain-Barr'e syndrome (P. =0.5). The mean age of patients with severe Guillain-Barr'e syndrome was less than of the subacute group but the difference is not statistically significant (P. =0.1).

Symptoms were preceded by an antecedent event in 77.3% of patients with severe Guillain-Barr'e syndrome and in 54.5% of the subacute (control) group .This difference is statistically significant (P=0.05) The majority of these events were URTI(68%) ,followed by GITI (4.5%) ,Tetanus toxoid containing(DPT) vaccination (4.5%)while 22.7% did not record any antecedent event. This is noted in other studies and the list of antecedent events is expanding to include rabies, polio, influenza, measles, and hepatitis- B vaccines [5] .In about 36.4% of patients with severe Guillain-Barr'e syndrome the illness was heralded by sensory symptoms (pain and parasthesia) and 22.7% of patients with subacute disease had these symptoms and the difference is not significant (P. = 0.3). Autonomic dysfunction was noted in 45.5% of cases manifested as either excess or reduced activity of the sympathetic or parasympathetic nervous system .Pulse and blood pressure changes were the commonest manifestations of dysautonomia, while only 27.3% of the control group had dysautonomia .The difference is statistically significant (P.= 0.03). Bulbar palsy was the commonest type of cranial nerve involvement,it was noted in 77.35% of cases and in 36.4% of the controls this difference is highly significant(P.= 0.003) this is in agreement with Nicolas et al who concluded that progression to mechanical ventilation was likely to occur in those patients with bulbar dysfunction [3]. The associated bulbar weakness and autonomic instability contribute to the necessity for control of the airways and ventilation [2].The mean SiO2 in patients with severe Guillain-Barr'e syndrome was 87.18 ± 5.65% and that in the subacute group was 96.3±2.71% The difference is statistically significant (P.0.03)and hence the decision to incubate which may be an anticipation of further deterioration and this indicates that the case at risk patients may be best managed in the intensive care unit .The mean CSF protein in patients with severe Guillain-Barr'e syndrome was 88.43 ± 66.47 mg/dl and that in subacute group is 88.42±69 mg/dl.The difference isn't statistically significant (P. = 0.5). As regard specific treatment 18 patients with severe Guillain-Barr'e syndrome were treated by IVIG and corticosteroid and 4(2 of them received partial parenteral nutrition) were treated by IVIG with out corticosteroid as it had not been suggested. A retrospective multicentre study found that IVIG accelerated recovery in children with Guillain-Barr'e syndrome who are unable to walk [10] .However a pilot study suggested that combination of treatment with IV methylprednisolone and IVIG for 5 days are more beneficial than IVIG alone [11] . The mean duration of mechanical ventilation in patients with severe Guillain-Barr'e syndrome that were treated by partial parentral nutrition was significantly less than similar cases who did not receive partial parentral nutrition, and this reflects the vital role of supportive treatment in artificially ventilated patients with severe Guillain-Barr'e syndrome as it may reduce the duration of mechanical ventilation and RICU stay by about 3 days.

In the present study the mean duration of mechanical ventilation was 12.5 ±53 days and the mean duration of stay in RICU was 25.85 ± 6.03 days ,and this is in agreement with large multicentre study where the mean time to reach improvement was 28 days[12]. Progression to mechanical ventilation was highly likely to occure in those patients with rapid disease progress (less than one week) bulbar dysfunction and dysautonomia, and low SiO2 at admission.

While inherently unpredictable, the course of patients with severe Guillain-Barr'e syndrome can to some extent be predicted on the bases of clinical information and simple bed side test may be used in the decision regarding admission to the RICU and preparation for elective intubations.

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