

Recovery of consciousness and life expectancy of children in a vegetative state

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The vegetative state does occur in children and is most commonly due to acquired traumatic and non-traumatic injuries. However, neurometabolic and degenerative diseases, as well as certain developmental brain malformations such as anencephaly, can also cause this condition. There are limited data available in children concerning recovery of consciousness and function from the vegetative state as well as life expectancy. This review concentrates on these issues and is based primarily on the data published in the Multi-Society Task Force Report on PVS which was published in 1994 as well as other epidemiological studies. Children in a vegetative state do have a poor prognosis for recovery of consciousness and function and do have a shortened life expectancy. Further research is needed to better understand what variables might contribute to recovery and what therapies might be of benefit.

INTRODUCTION

Very limited data are available regarding recovery from the vegetative state (VS) and the life expectancy of children with this condition. In 1994, the Multi-Society Task Force (MSTF) on the persistent vegetative state published data extracted from the world's literature regarding these two issues (MSTF, 1994) and since then several epidemiological studies have been published that have further examined these issues (Ashwal, Eyman, & Call, 1994; Strauss, Shavelle, & Ashwal, 1999; Strauss, Ashwal, Day, & Shavelle, 2000).

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TABLE 1
Worldwide prevalence of children aged less than 15 years in a vegetative state

<i>Region</i>	<i>Total Population (<.15 years)</i>	<i>Average number of VS patients</i>	<i>Low estimate of number of VS patients</i>	<i>High estimate of number of VS patients</i>
World	1,894,200,000	92,816	11,365	151,536
Continents				
North America	67,830,000	3,324	407	5,426
Latin America	172,800,000	8,467	1,037	13,824
Europe	123,590,000	6,056	742	9,887
Asia	1,149,000,000	56,301	6,894	91,920
Africa	361,620,000	17,719	2,170	28,930
Selected Countries				
Belgium	1,872,000	92	11	150
China	283,514,000	13,892	1,701	22,681
Denmark	1,026,000	50	6	82
Finland	936,000	46	6	75
France	11,362,000	557	68	909
Germany	12,390,000	607	74	991
Greece	1,540,000	75	9	123
Iceland	69,000	3	0	6
India	384,696,000	18,850	2,308	30,776
Ireland	840,000	41	5	67
Israel	1,876,000	92	11	150
Italy	8,008,000	392	48	641
Japan	17,850,000	875	107	1,428
Netherlands	3,078,000	151	18	246
Norway	920,000	45	6	74
Portugal	1,664,000	82	10	133
Russia	26,190,000	1,283	157	2,095
Spain	6,195,000	304	37	496
Sweden	1,620,000	79	10	130
Switzerland	1,241,000	61	7	99
United Kingdom	11,248,000	551	67	900
United States	61,215,000	3,000	367	4,897

Population data obtained from <http://www.prb.org/datafind/datafinder.htm>. Prevalence rates are based on previously published data as summarised in Ashwal et al., 1994. These prevalence data were then used to calculate an average prevalence rate and this rate was used for the population of children <15 years of age for each region or country listed in the table. Overall, the average prevalence of VS patients was 49 per million with the low estimate being 6 per million and the high estimate being 80 per million.

EPIDEMIOLOGY

It is difficult to obtain accurate data regarding the prevalence of VS among children. Based on previously published estimates from many countries and available worldwide population data, it is possible to estimate the number of children in a VS (Table 1). The estimated number of children aged

under 15 years worldwide in a VS is approximately 93,000 (range 11,365–151,536).

As the neurological insults occurring in children are different from adults, the risk for developing as well as recovering from VS also differs (Ashwal et al., 1994). Acute traumatic and non-traumatic injuries to the nervous system in children accounts for approximately 30% of cases. Perinatal insults (17.7%), chromosomal disorders or congenital malformations (13.0%), and infections (10.3%) occur less frequently (Ashwal et al., 1994). As clinical experience suggests, in a number of patients (28%), no specific cause can be determined. The aetiology of VS in children can be classified into three broad groups of disorders including (1) acute traumatic and non-traumatic brain injuries; (2) metabolic and degenerative disorders affecting the nervous system; and (3) developmental malformations. Children who are in VS due to acquired brain injury, particularly if it is traumatic, have the best chance for recovery of consciousness and function whereas children in whom the aetiology is due to a metabolic or degenerative disease are unlikely to recover consciousness and may have a shorter life expectancy (MSTF, 1994). In children in whom the VS is due to a cerebral malformation, the chances of recovery of consciousness remain small, but long-term studies of such individuals have not been reported.

RECOVERY

As described in the MSTF report, recovery from VS can be considered in terms of recovery of consciousness and recovery of function (MSTF, 1994).

Acute traumatic and non-traumatic injuries in children

Traumatic injuries. Recovery of awareness from post-traumatic VS appears to be somewhat better in children compared to adults. The MSTF on PVS has collected data (Table 2) on the potential for recovery from VS after severe traumatic brain injury in adults and children. Of 106 children in VS one month after severe head injury, 24% regained awareness by 3 months. At 1 year, 29% remained in VS, 9% had died, and 62% had recovered consciousness. Late recoveries after 12 months were not reported although a study by Kriel and co-investigators found that two of 40 children with traumatic brain injury began to recover after 1 year in VS (Kriel, Krach, & Jones-Saete, 1993). One patient had limited language function and was described as "able to express wants and needs". The other child had no language but was socially responsive and smiled in response to a voice or face. It is not clear whether this patient actually regained consciousness. In this study, eight of nine patients in VS for less than 3 months recovered to a severe disability; the remaining

TABLE 2
Incidence of recovery of consciousness and function in children in VS one month
after traumatic and non-traumatic brain injury

	Outcome at 3, 6, and 12 months as a percentage of children diagnosed VS 1 month after insult			Functional recovery of those patients who recovered consciousness by 12 months	
	3 months (%)	6 months (%)	12 months (%)	Recovery	(%)
Traumatic (n = 106)					
Dead	4	9	9	Severe disability	35
VS	72	40	29	Moderate disability	16
Recovered consciousness	24	51	62	Good recovery	11
Total	100%	100%	100%		62%
Non-traumatic (n = 45)					
Dead	20	22	22	Severe disability	7
VS	69	67	65	Moderate disability	0
Recovered consciousness	11	13	13	Good recovery	6
Total	100%	100%	100%		13%

This table was adapted from the Multi-Society Task Force on PVS (1994).

patient was moderately disabled. In the 15 patients in VS for 3–6 months, 12 recovered to a severe disability and three to a moderate disability and in those five patients in VS for 6–12 months three were severely and two moderately disabled. No good recoveries were reported in this series of 40 children who were in post-traumatic VS for 3 months. This data can be compared to the MSTF report where it was observed that of the 62% of children who did recover consciousness from post-traumatic VS, recovery of function (based on the use of the Glasgow Outcome Scale score) were: good recovery (11%), recovery to a moderate disability (16%), and recovery to a severe disability (35%).

Only one other study has provided data on recovery after TBI in children who were in VS one month after injury (Heindl & Laub, 1996). At 19 months post-injury, 84% of children (total $n = 82$) had recovered consciousness and 16% of these children became independent in daily activities. Less than 5% of children recovered consciousness beyond nine months of injury.

Non-traumatic injuries. Children in non-traumatic VS have a much poorer potential for recovery of consciousness and function at 12 months post-injury than from traumatic VS (MSTF, 1994). Data collected by the MSTF on PVS showed that only 11% of children regained awareness by 3 months after injury (MSTF, 1994). At 1 year most children remained in VS (65%) or died (22%); only 13% showed recovery and this was usually to a severe disability.

Good or moderate functional recovery is extremely unlikely but may occur in children in VS due to a non-traumatic brain insult. In most of the patients who show recovery, awareness can usually be detected within 3 months of injury. Kriel et al. also reported in their recent studies that three of 13 children in post-anoxic VS for longer than 12 months (i.e., 370, 480, and 840 days) showed recovery of consciousness (Kriel et al., 1993). These patients recovered to a severe disability and in some of these children it was unclear whether this reflected recovery of consciousness or a patterned subcortical behavioural response.

Additional data from another study on post-hypoxic VS ($n = 55$) found that only 55% recovered consciousness by 19 months and only 4% became independent (Heindl & Laub, 1996). Less than 5% recovered beyond 9 months after injury. In this study children in VS due to hypoxic ischaemia also had a higher incidence of seizures and complications such as pneumonia, gastrointestinal complications and heterotopic calcifications.

Degenerative and metabolic disorders

Children in VS due to degenerative or metabolic diseases have virtually no possibility of recovering because these diseases are progressive or reach a clinical plateau in their terminal stages. In some children who are not vegetative but severely disabled an intercurrent illness may cause them to appear vegetative. As the illness improves the child may recover to his/her previous state of limited cognition.

Developmental malformations

Infants and children with congenital brain malformations severe enough to cause developmental VS are unlikely to acquire awareness. Anencephaly is the only malformation in which the prognosis for no recovery can be made at birth (Medical Task Force on Anencephaly, 1990). Other malformations diagnosed at birth may result in a vegetative outcome and if confirmed by clinical examination at 3–6 months of age, the prognosis for any improvement is extraordinarily small. The majority of infants with such malformations who recover consciousness have extremely limited awareness and minimal functional capacities.

Probability for recovery

The MSTF on PVS (1994) has estimated the probability for recovery of consciousness and function from traumatic and non-traumatic VS in adults and children who were vegetative one month after an acute injury (Table 3). Calculations of the probability for recovery were based on data from the

TABLE 3
 Probabilities for recovery of consciousness and function at 12 months in children in PVS after traumatic and non-traumatic brain injury

	<i>Outcome probabilities at 12 months</i>	
	<i>Traumatic PVS (n = 106)</i>	<i>Non-traumatic PVS (n = 45)</i>
<i>Patients in PVS for 3 months</i>		
Dead (%)	14	3
PVS (%)	30	94
Severe (%)	24	3
Mod/Good (%)	32	0
<i>Patients in PVS for 6 months</i>		
Dead (%)	14	0
PVS (%)	54	97
Severe (%)	21	3
Mod/Good (%)	11	0

Modified from the Multi-Society Task Force on PVS (1994). Severe = recovery to a severe disability; Mod/Good = recovery to a moderate disability or to a good recovery.

previous sections, which provided the actual frequencies of recovery. Outcome probabilities at 12 months were determined for patients who remained vegetative at 3 and 6 months. Functional recovery was determined for two possible outcomes: (1) good recovery or recovery to a moderate disability and (2) recovery to a severe disability. Based on these probabilities (MSTF, 1994), the following statements can be made:

1. After 3 months, children in post-traumatic VS have a 56% chance of recovering consciousness in contrast to only 3% in children in non-traumatic VS. Of those children who recover, the probability for recovering to a severe disability is 24% and of making a moderate or good recovery 32%.
2. After 6 months, children in post-traumatic VS have a 31% chance of recovering consciousness in contrast to only 3% of children in non-traumatic VS. However, the chance of making a moderate or good recovery is now much less (i.e., 11%); recovery to a severe disability is more likely (21%).
3. VS is also likely to be permanent 12 months after traumatic brain injury and 3 months after non-traumatic injury in children. The chance for recovery after this time period seems to be exceedingly rare and almost always to a severe disability.

Life expectancy

Both adults and children in VS have shortened life expectancies despite preservation of brain stem and autonomic functions. In adults, long-term studies have shown that about 82% of patients will die within 3 years and about 10% of adults who are in VS will survive 5–10 years and only 4% beyond that time (MSTF, 1994).

As noted in Table 2, 91% of children in VS one month after traumatic brain injury were alive at one year; of those children in VS from non-traumatic injury, 78% survived. A large population-based study examining 847 children and adults considered to be in VS found approximately the same duration of survival for older children but a much shortened median life expectancy in children aged under 1 year (Ashwal et al., 1994). The life expectancy of infants and children in VS appeared to be an age-dependent phenomenon. For example, the median survival time of children less than 1 year of age was 2.6 years in contrast to children aged 2–6 years where it was 5.2 years. There is also likely to be some relation between certain aetiologies of the VS and survival times. For the data available it appears that children in VS from non-traumatic injury (8.6 years) and chromosomal disorders (8.2 years) have a longer life expectancy than children in whom the VS is due to perinatal disorders (4.1 years), traumatic brain injury (3.0 years), or infection (2.6 years). This does not appear to be due to any interdependency between age and aetiology. The reasons for these limited differences remain unclear. One possible explanation is that the shortened life expectancies observed in the perinatal or infection groups may be due to a greater degree of permanent brain stem or hypothalamic injury which causes greater immobility, an increased risk for aspiration, and an overall poorer nutritional status which renders these patients more susceptible to infection or cardiorespiratory arrest.

Life expectancy does not appear to be affected by the residential location of a child in VS. There were no differences reported in life expectancy in those children living at home (4.5 years) compared to those living in an institution (5.2 years); life expectancy of children living in a skilled nursing facility or private hospital setting was somewhat shortened (3.2 years). Although there is no definite reason to explain these limited differences in life expectancy, our experience suggests that the associated medical problems of VS patients are similar irrespective of facility. In cases where there are more serious acute or chronic medical problems, additional home nursing care is frequently provided to families or the incidence of hospital readmissions is increased; if patients are in institutions or skilled nursing facilities their care levels are upgraded to meet their specific needs.

Additional data have been reported concerning life expectancy of children in a permanent vegetative state (Strauss et al., 1999) as well as comparing life expectancy of children in VS to those in a minimally conscious state (MCS)

(Strauss et al., 2000). Both studies were from a similar population as the above-cited paper (i.e., Ashwal et al., 1994) that was based on data from patients who were residents of California and in whom an annual Client Development Evaluation report was completed. The first report was based on data from 1,021 VS patients and two major findings emerged. The first was that of a secular trend towards higher life expectancy in more recently diagnosed VS patients (Strauss et al., 1999). Life expectancy was 3.6 years in VS patients diagnosed in 1980 compared to 7.2 years for those diagnosed in 1990. Also life expectancy was longer in patients who were in VS for 4 years (life expectancy of 12.2 years) compared to those who were in VS for 1 year (10.5 years). In the second study (Strauss et al., 2000), no differences were found in the percentage of patients surviving for 8 years between those patients in VS (63%) and those in an immobile MCS (65%) or a mobile MCS (81%). These findings suggested that the presence or absence of consciousness might not be a critical factor determining life expectancy.

There are no data concerning extraordinary long survival (i.e., greater than 15 years) for children in VS. Rare cases of prolonged survival for periods up to 10–20 years were reported in the survey of child neurologists (Ashwal, Bale, Coulter, Eiben, Garg, Hill et al., 1992). The MSTF on PVS (1994) has estimated that the probability of an individual patient having prolonged survival is less than 1 in 15,000 to 75,000.

REFERENCES

- Ashwal, S., Eyman, R. K., & Call, T. L. (1994). Life expectancy of children in a persistent vegetative state. *Pediatric Neurology, 10*, 27–33.
- Ashwal, S., Bale, J. F., Coulter, D. L., Eiben, R., Garg, B. P., Hill, A., et al. (1992). The persistent vegetative state in children: Report of the Child Neurology Society Ethics Committee. *Annals of Neurology, 32*, 570–576.
- Heindel, U. T., & Laub, M. C. (1996). Outcome of persistent vegetative state following hypoxic or traumatic brain injury in children and adolescents. *Neuropediatrics, 27*, 94–100.
- Kriel, R. L., Krach, L. E., & Jones-Saete, C. (1993). Outcome of children with prolonged unconsciousness and vegetative states. *Pediatric Neurology, 9*, 362–368.
- Strauss, D. J., Ashwal, S., Day, S. M., & Shavelle, R. M. (2000). Life expectancy of children in vegetative and minimally conscious states. *Pediatric Neurology, 23*, 312–319.
- Strauss, D. J., Shavelle, R. M., & Ashwal, S. (1999). Life expectancy and median survival time in the permanent vegetative state. *Pediatric Neurology, 21*, 626–631.
- Multi-Society Task Force Report on PVS (1994). Medical aspects of the persistent vegetative state. *New England Journal of Medicine, 330*, 1499–1508, 1572–1579.
- Medical Task Force on Anencephaly (1990). The infant with anencephaly. *New England Journal of Medicine, 322*, 669–674.