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Sub-Acute Sclerosing Panencephalitis at University Hospital of Constantine

Bouhdjila A[1], Benhacine Z[2]

[1]Faculté de medecine université constantine3 Algerie ;*Service de pédiatrie Centre hospitalo-universitaire de Constantine

[2]Faculté de médecine ,Constantine3 université SB Service de pédiatrie B , chu de Constantine

ABSTRACT

Introduction Subacute sclerosing panencephalitis (SSPE) is a serious, late complication of measles, and the prognosis remains poor in the absence of effective treatment, underscoring the role of vaccination in its prevention.

Objectives We describe the clinicobiological and evolutionary profile of 16 patients recruited from the B chu de Constantine pediatric department.

Patients and methods Study of16 cases: epidemiological, clinical, EEG, biological and evolutionary profile.

Results16 cases of SSPE were collected, 75% were male, 70% were aged between 24 and 59 months, almost all patients had had measles, 98% within the first 24 months of life, 93% had not been vaccinated against measles, SSPE occurred in more than 50% of cases between 24 and 48 months after measles, all had received treatment (100% corticosteroids and immunoglobulins), 87% antiepileptics and 25% rituximab, the course was characterized by stabilization: 27%, death: 33% deterioration:40% of cases.

Comments and discussion SSPE is a rare complication occurring a few years after recovery from measles Almost all our patients had not been vaccinated against measles.

in our patients, the median age at onset of SSPE was 4.5 years This earlier onset of SSPE in recent studies than reported in older studies suggests early onset of measles, viral mutation or a fragile immune system.

Almost all patients were unvaccinated against measles; 40% of patients died, 31% worsened (stage IV) and 37% stabilized.

Conclusion Given the poor prognosis of SSPE, the best way to control this disease is to vaccinate the target population against measles.

KEYWORD : measles, SSPE , prognosis ,vaccination, prevention.

ARTICLE DETAILS

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In 2018, a measles epidemic arose in Algeria following a refusal of vaccination by the population in 2017. This massive refusal led to the outbreak of this epidemic and ended in a significant morbidity and mortality, three years after this health event we are currently witnessing the appearance of a very serious encephalopathy in a population aged 3 to 5 years these children have all been victims of the measles disease and were not vaccinated against measles. This is subacute sclerosing panencephalitis (PESS), which has rarely been seen in Algeria since the introduction of the measles vaccine in the 80s, which further emphasizes the major role of vaccination in the fight against measles and its harmful consequences.

History

Subacute sclerosing pan encephalitis (SESS) is a rare but serious neurodegenerative disease due to the often dramatic outcome leading either to death or to a severe sensorineural disability.It is cited for the 1st time in 1945 by Von Bogart describing the pathological lesions on the biopsy of the brain of a 13-year-old child who died by this pathology [1] .It was recognized as a clinical and anatomopathological syndrome by Jabbour in 1972, The latter established a national register to identify these cases. [2]. Between 1968 and 1971, Mailler and then Connolly provided the biological proof, the 1st discovered anti-measles antibodies in the CSF of 10 patients with PESS.In 1990 Brimani described its EEG characteristics [3,4]

The great variability of the clinical symptomatology, the electroencephalography aspect, the evolution, make subacute sclerosing panencephalitis a disease that remains little known The therapeutic possibilities are very limited due to this misunderstanding and the outcome is often fatal

isoprinosine monotherapy and isoprinosine plus ribavirin were the standard treatments, but also intravenous immunoglobulin therapy, intrathecal a-interferon (a-IFN), and amantadine therapy

upto now results not show no satisfaction the vaccination is the only mean that will eradicates measles and its harmful consequences [14]

Objectives

We report the clinicobiological and evolutionary profile of a series of 16 cases managed during the year 2021 and 2022 at the pediatric service B chu of Constantine

Patients and methods

Study of files of 16 children: epidemiological profile; EEG and biological clinic and evolution of PESS. Retrospective study over 2 years from January 2020 to March 2022.

The diagnosis of PESS is based on three inseparable criteria: clinical, electrophysiological, and biological.

The EEG may be normal at the beginning and does not eliminate the dgc interest in repeating in 15 days +specific tests hyperpnea test intermittent light stimulation background periodic complexes appear and the myoclonus that sometimes accompany them succeed them in 4-8months ICD test favors the reappearance of periodic complexes and the disappearance of myoclonus. [7]

At the beginning of the PESS the periodicity is fast 10 seconds between 2 complexes; during evolution it becomes slow up to 15 seconds and more: Periodic slow waves short periodicity1 to 2 seconds.[8]

The intrathecal synthesis of anti-measles virus antibodies in patients needs to be evaluated. It is she who differentiates PESS from other neurological complications of measles: acute encephalitis with inclusions and post-infectious encephalitis.[9]

To define the case of PESS it is necessary to bring together at least one major criterion and one or more minor criteria] the clinical picture is presented in several forms depending on the evolutionary stage [ref] which ranges from the decline in cognitive functions to the state of decerebration (see table below)

DIAGNOSIS OF PESS

It takes a major criterion plus one or more minor criteria (See table below)

Major criterion

-1 Disorders of consciousness for more than 24 hours

- 2 Ataxia

- 3 Behavioral disorders

Minor criterion

Table1. Diagnosis of SSPE

Major criteria	Minor criteria
- Disorders of consciousness for more than 24 hours - Ataxia - Behavioral disorders	1. Fever 2. Convulsions 3. Signs of focalizations 4. CSF Abnormalities 5. EEG Abnormalities 6. Brain MRI abnormalities

Table 2. Evolutionary stages of PESS

stage	Clinical features
Stage I	Intellectual deterioration and behavioral disorders with decreased academic performance and agnosia, apraxia, attentional deficits, distractibility
Stage II	Myclonies, incoordination choro-athetosis
Stage III	Extrapyramidal dysfunction, opisthotonos
Stage IV	Respiratory irregularity, increasing hypotonia, decerebration rigidity, autonomic dysfunction

RESULTS

16 cases of subacute sclerosing pan encephalitis are collected, all meeting the definition of PESS at different evolutionary stages of the disease, the socio-demographic and clinical characteristics are reported on Table III

ANALYSIS OF THE RESULTS

Among our patients, 75% are male with a sex ratio of 2, 70% are aged 24-59months and 12% aged less than 24months.

For measles all patients have had measles and in 98% in the first 24 months of their life ,93% have not been vaccinated against measles

PESS occurred in more than 50% of cases between 24 and 48 months after the onset of measles

All patients have a disturbed EEG with 67% of PLOS and 22% a paroxysmal trace 11% an aspect of delta brush

Clinically, only 6% are at stage I of the disease, 50% are at stage II 14% stage III 31% stage IV

Biology is disturbed in all cases with in 50% presence of oligoclonal peak of anti-measles antibodies in CSF, presence of anti-measles antibodies in the blood witnesses of measles disease and pleiocytosis in 20% of cases

Therapeutically, all have benefited from pharmacotherapy (100% corticosteroid therapy and immunoglobulins), anti epileptics87% and rituximab in 25%

The evolution was characterized by stabilization in 27%, in 33% and clinical deterioration in 40% of cases.

COMMENTS AND DISCUSSION

Measles, once a killer of children, has become a mild infection that heals in a few days in the era of vaccination vaccination. The virus can take refuge in the cns latently and remaining for several years until its reactivation during the decrease in immunity to trigger the dreaded pan encephalitis this complication is rare occurs a few years after the cure of measles (1/1400 before 5 years and 1/600 before 12 months.) occurring on average 10 years after measles.[9,10,11]

Almost all of our patients were not vaccinated against measles except one who was vaccinated at 12 months and rash at 15 months and PESS at 24 months)

in our patients the median age of occurrence of PESS is 4.5A This precocity of occurrence of PESS in recent studies compared to what has been reported in old studies suggests the early occurrence of measles or viral mutation.[8]

A male predominance is noted 3/1. As in all studies [11]

almost all of our patients had measles, as in all studies with a variable degree of 60 to 78% before 2 years [2,10] confirming the hypothesis that PESS is a late complication of measles.

The interval between the occurrence of measles and the onset of pess is 3.5 A joining the other studies and it is later in the older studies 5.6years 11]

Children vaccinated for measles have less severe forms of PEES (only one case of our patients) 5 non-vaccinated cases in Tanya's series [8]

As has been observed in England, France or Georgia, the number of cases of PESS has increased and there have recently been 16 cases between 2008 and 2017 due to a decrease in vaccination coverage [12]

Almost all of our patients were unvaccinated against measles

EVOLUTION

We had the death of 40% of our patients, the aggravation (stage IV) of 31% and a stabilization of 37% approaching the English series of Tanya Lam 30% [8]

Vaccination minimizes infection with the measles virus and protects against pestilence, in the case of transient immunodeficiency. Vaccination with an attenuated strain of the measles virus is safe for immunocompetent children. [9]

The prognosis of PESS being poor, the only way to control this disease is vaccinationin fact The number of patients with PESS is positively correlated with the number of patients with measles and the incidence of PESS is inversely proportional to the rate of vaccination against measles. This has been demonstrated in high-income countries where the prevalence of MS has been steadily declining since the introduction of measles vaccination in the 1960s. [13]

CONCLUSION AND PROSPECTS

PESS is a rare but fatal complication of measles in children

A national register of measles cases and their medium and long-term follow-up must be established to measure the extent of the long-term consequences of measles.

Only mass vaccination with a good coverage exceeding 95% of the population can prevent it as long as the

Table 3. biological ,EEGand MRI results

Examen Résultats	n	%
LCR		
Normal	13	81
Lymphocytose	1	6
Hyperproteinorachie sup 1g	2	12
IRM CEREBRAL		
N	11	75
Hyper signal sub blanche PV	4	25

EEG		
OL PERIODIQUE OU PSEUDOP	12	87
DELTA BRUSH	2	12
ACTIVITE PAROXYSTIQUE	4	25
POC+		
0,72-1	1	6
1-2	2	12
3-4	3	19
4-10	3	19

POC pic oligoclonal des anticorps anti rougeole
OL onde lente
PV periventriculaire

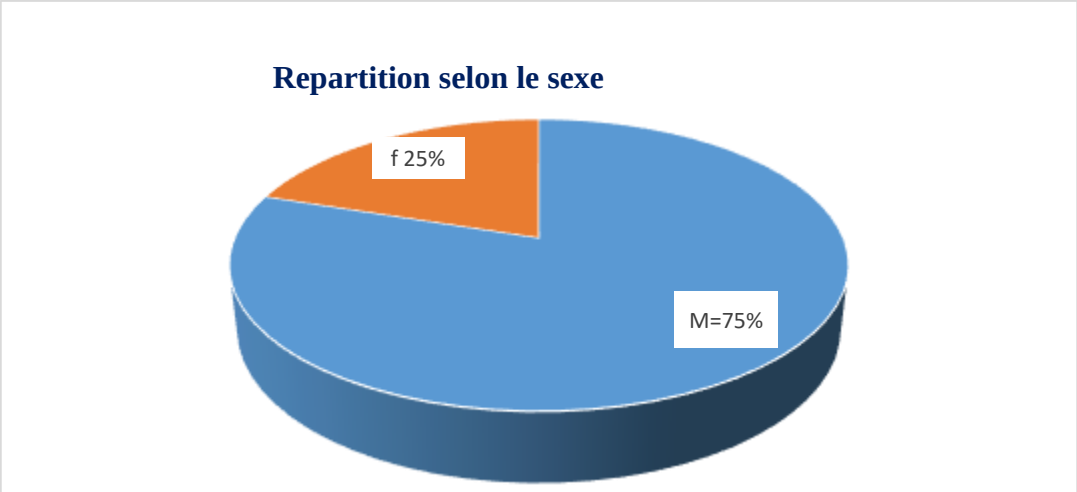

Fig. 1. Répartition according of the gender

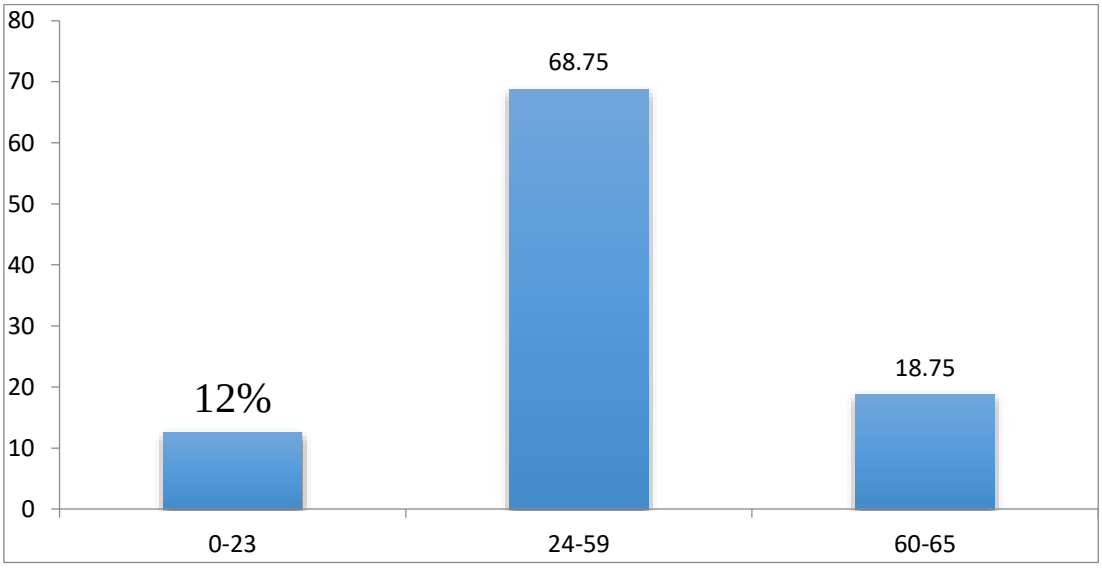

Fig. 2. Repartition according of the age

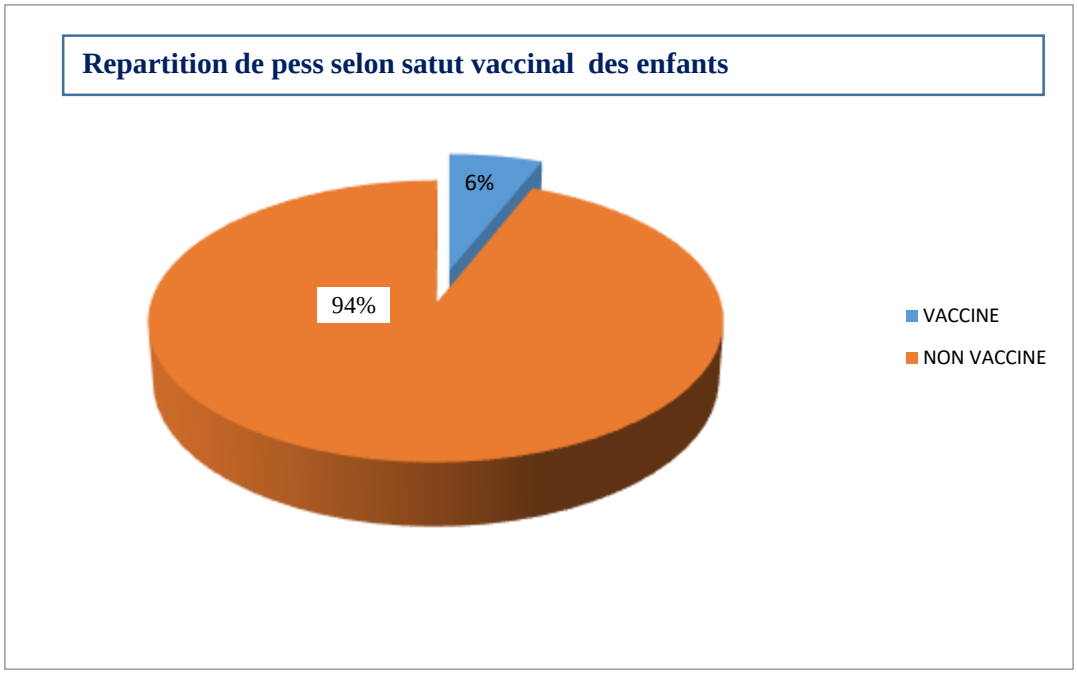

Fig. 3. Repartition according of the vaccinal status

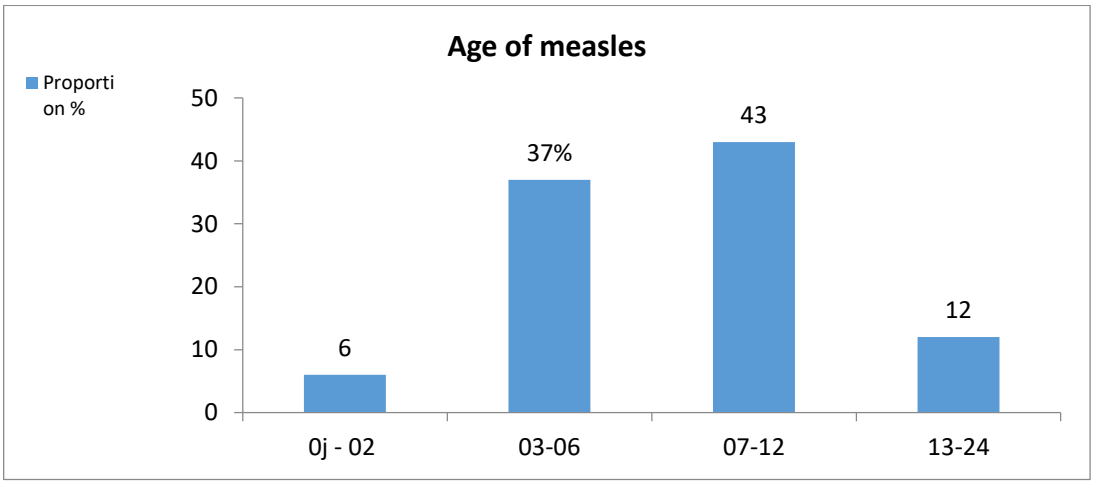

Fig. 4. Répartition according of the age starting of measles

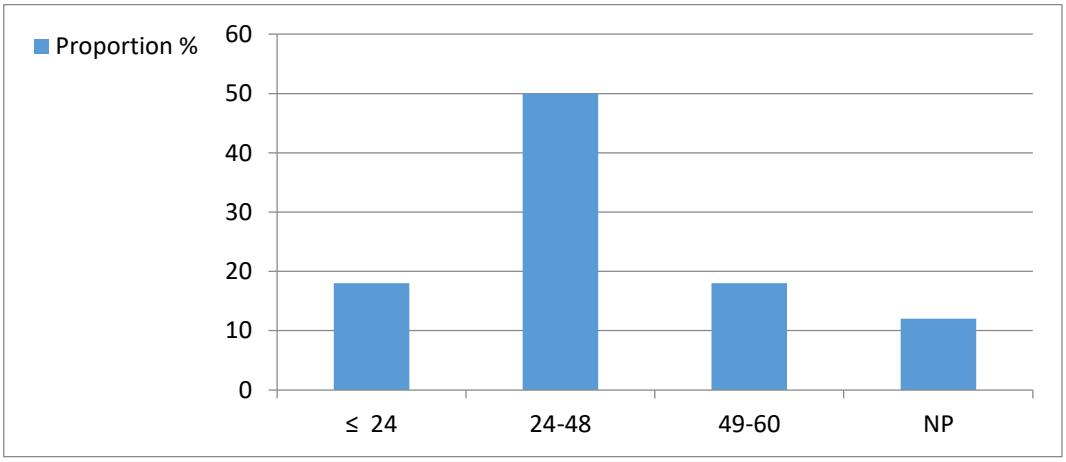

Fig. 5. delay between measles and PESS symptoms

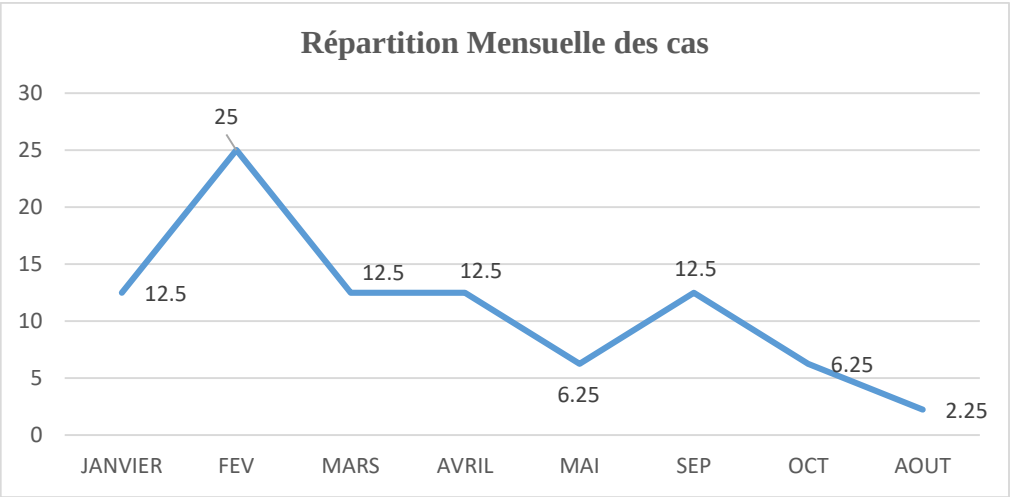

Fig. 6. monthly repartition of cases

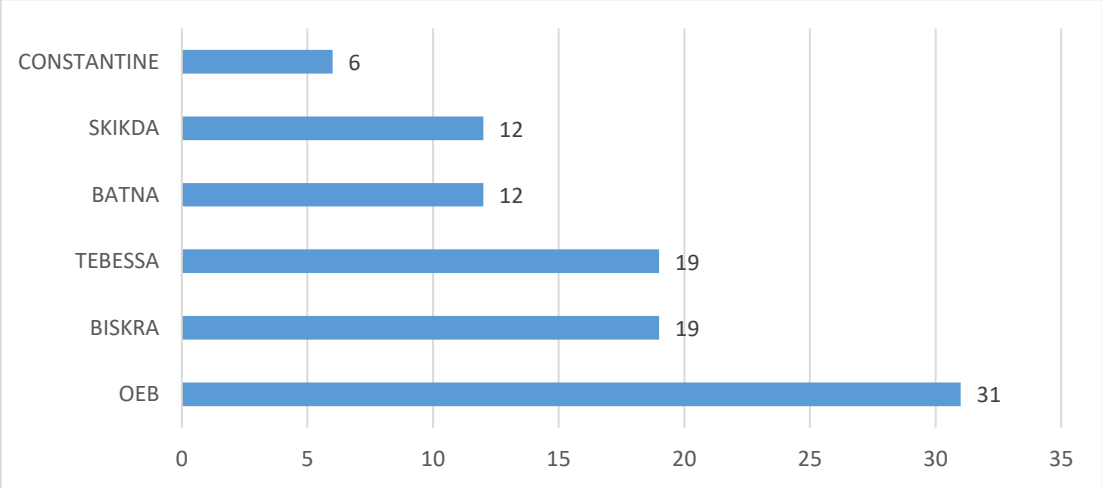

Fig. 7. Distribution according to the origin of patients

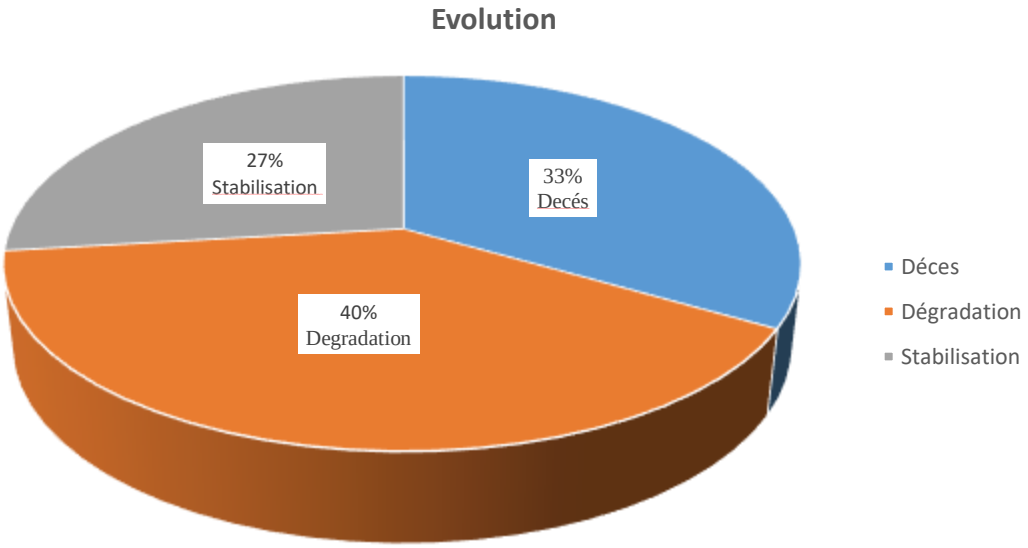

Fig. 8. Distribution according to the evolution of patients

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