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29.30
janvier
2026

19^e CONGRÈS
DE LA SOCIÉTÉ FRANÇAISE
D'HÉMAPHÉRÈSE

LES TERRASSES DU PARC, 115 BOULEVARD STALINGRAD
LYON - Villeurbanne



Efficacité et tolérance de l'aphérèse thérapeutique en neuropédiatrie

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UNIVERSITÉ
DE MONTPELLIER

ORKid
FILIERE ORPHAN
KIDNEY
DISEASES

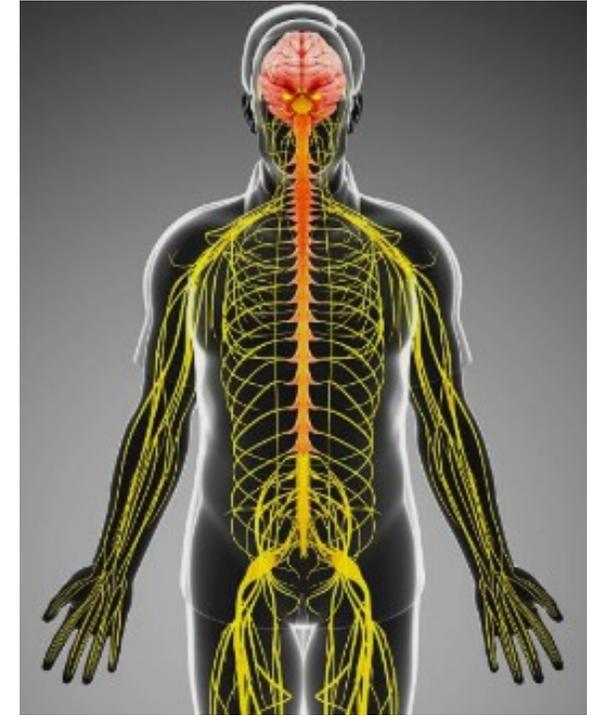


Liens d'intérêt

- Pas de liens d'intérêt
- ... IA free!

Pathologies neurologiques inflammatoires accessibles aux TPE/IA

- Système nerveux central:
 - encéphalites/ encephalomyélites auto-immune
 - Myélite transverse
 - ADEM
 - Sclérose en plaque
 - NMOSD
 - MOGAD
 - Encephalite avec Ac anti NMDA
- Système nerveux périphérique:
 - Guillain–Barre syndrome (GBS) avec sous types
 - Polyradiculonevrite inflammatoire demyelinisante chronique (PIDC)
- Jonction neuromusculaire: myasthenie



Incidence des atteintes neurologiques immunes dans la population pédiatrique

- GBS : 0.5-1.5 / 100000
 - acute inflammatory polyradiculoneuropathy (AIDP): 0.34-1.5/100000
 - acute motor axonal neuropathy (AMAN): 0,03-0.09/100000
 - acute motor-sensory axonal neuropathy (AMSAN) 0.03-0.05/100000
 - Miller-Fisher syndrome 1/1000000
- Myasthenia gravis:
 - Transient neonatal myasthenia gravis
 - Juvenile myasthenia gravis; 5.9-8.7/1000000
- Pediatric onset multiple sclerosis: 0.5/100000
- PICD: 0.3/1000000
- Encephalite de Rasmussen 0.15-0.24/1000000
- NMOSD: 0.31/1000000
- Lambert Eaton syndrome <50 cas décrits

DiFazio et al. Medscape 2025
Ipe et al. Frontiers Neurology. 2021
Ipe et al J Clin Apher 2021

Indications IA/EP en neurologie: ASFA guidelines 2016

Disease	TA modality	Category
Acute disseminated encephalomyelitis	TPE	II
Acute inflammatory demyelinating polyradiculoneuropathy/Guillain-Barré syndrome	TPE	I
Chronic inflammatory demyelinating polyradiculoneuropathy	TPE	I
Dermatomyositis/polymyositis	TPE	IV
Hashimoto's encephalopathy: Steroidresponsive encephalopathy associated with autoimmune thyroiditis	TPE	II
Lambert-Eaton myasthenic syndrome	TPE	II
Multiple sclerosis	TPE	II
	IA	III
Myasthenia gravis	TPE	III
	TPE	I
Neuromyelitis optica spectrum disorders	TPE	I
	TPE	II
N-methyl D-aspartate receptor antibody encephalitis	TPE	III
Paraneoplastic neurological syndromes	TPE	I
	IA	III
Paraproteinemic demyelinating neuropathies/chronic acquired demyelinating polyneuropathies	TPE	III
	TPE	IV
	TPE	I
	TPE	I
Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections; Sydenham's chorea	TPE	III
	IA	III
	TPE	II
Progressive multifocal leukoencephalopathy associated with natalizumab	TPE	III
Stiff-person syndrome	TPE	I
Voltage-gated potassium channel antibodies	TPE	III

I : traitement 1ère ligne
 II : traitement 2ème ligne
 III : efficacité non établie
 IV : inefficace / dangereux

Indications IA/EP en neurologie: ASFA guidelines 2023

TABLE 1 Category and grade recommendations for therapeutic apheresis.

Disease/condition	Indication	Procedure	Category	Grade	
Acute disseminated encephalomyelitis	Steroid refractory	TPE	II	2C	
Acute inflammatory demyelinating polyradiculoneuropathy	Primary treatment	TPE	I	1A	
		IA	I	1B	
Chronic acquired demyelinating polyneuropathies	IgG/IgA/IgM related	TPE	I	1B	
		Anti-myelin-associated glycoprotein	TPE	III	1C
		CANOMAD/CANDA ^a	TPE	III	2C
Chronic focal encephalitis		TPE/IA	III	2C	
Chronic inflammatory demyelinating polyradiculoneuropathy		TPE/IA	I	1B	
Idiopathic inflammatory myopathies ^a	Anti-synthetase-syndrome	TPE	III	2B	
	Clinically amyopathic dermatomyositis	TPE	III	2B	
	Immune-mediated necrotizing myopathies	TPE	III	2B	
Lambert-Eaton myasthenic syndrome		TPE	II	2C	
Multiple sclerosis	Acute attack/relapse	TPE	II	1A	
		IA	II	1B	
	Chronic primary or secondary progressive	TPE/IA	III	2B	
Myasthenia gravis	Acute, short-term treatment	TPE/DFPP/IA	I	1B	
	Long-term treatment	TPE/DFPP/IA	II	2B	
N-methyl-D-aspartate receptor antibody encephalitis		TPE/IA	I	1C	
Paraneoplastic autoimmune retinopathies ^a		TPE	III	2C	
Paraneoplastic neurological syndromes		TPE/IA	III	2C	
Pediatric autoimmune neuropsychiatric disorders	PANDAS/PANS, exacerbation	TPE	II	1B	
	Sydenham's chorea, severe	TPE	III	2B	
Progressive multifocal leukoencephalopathy associated with natalizumab		TPE	III	1C	
Steroid-responsive encephalopathy associated with autoimmune thyroiditis		TPE	II	2C	
Stiff-person syndrome		TPE	III	2C	
Sudden sensorineural hearing loss		LA/DFPP/TPE	III	2A	

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Clinical diagnosis of a confirmed autoimmune condition in which TPE may be of benefit

Moderate/severe presentation of peripheral nervous system disease:^a

- Chronic inflammatory demyelinating polyradiculoneuropathy
- Guillain-Barré syndrome
- Myasthenia gravis (including preoperative optimization)

Consider TPE as initial therapy

Severe/fulminant presentation of central nervous system disease:^b

- Acute disseminated encephalomyelitis/acute transverse myelitis
- Limbic encephalitis
- Multiple sclerosis
- Neuromyelitis optica spectrum disorders
- *N*-methyl-D-aspartate receptor antibody encephalitis

Inadequate response to steroid and/or IVIG after 48–72h^{c,d}

Consider TPE as rescue therapy

Preparation for TPE:

- Early identification of appropriate setting (usually HDU/intensive care)
- Early discussion with appropriate team with experience of delivering TPE in children
- Baseline bloods: albumin, calcium, U&E, FBC, clotting profile, blood group, hepatitis B/C, HIV, save serum
- Plan for central venous catheter insertion if required^e

Discussion of risks and benefits with family

Delivery of a course of TPE:

- Five to seven procedures of 1–1.5 total plasma volume exchanges on alternate days, unless otherwise stated in Table I
- Discussion with pharmacist regarding alterations to dosing of regular medications; continue adjuvant high-dose steroids if already commenced
- Premedication with paracetamol and antihistamine;^f involve a play specialist for younger children^g
- Close monitoring of vital signs and laboratory parameters

Adequate response: consider initiating other immunotherapies post stabilization with TPE^d

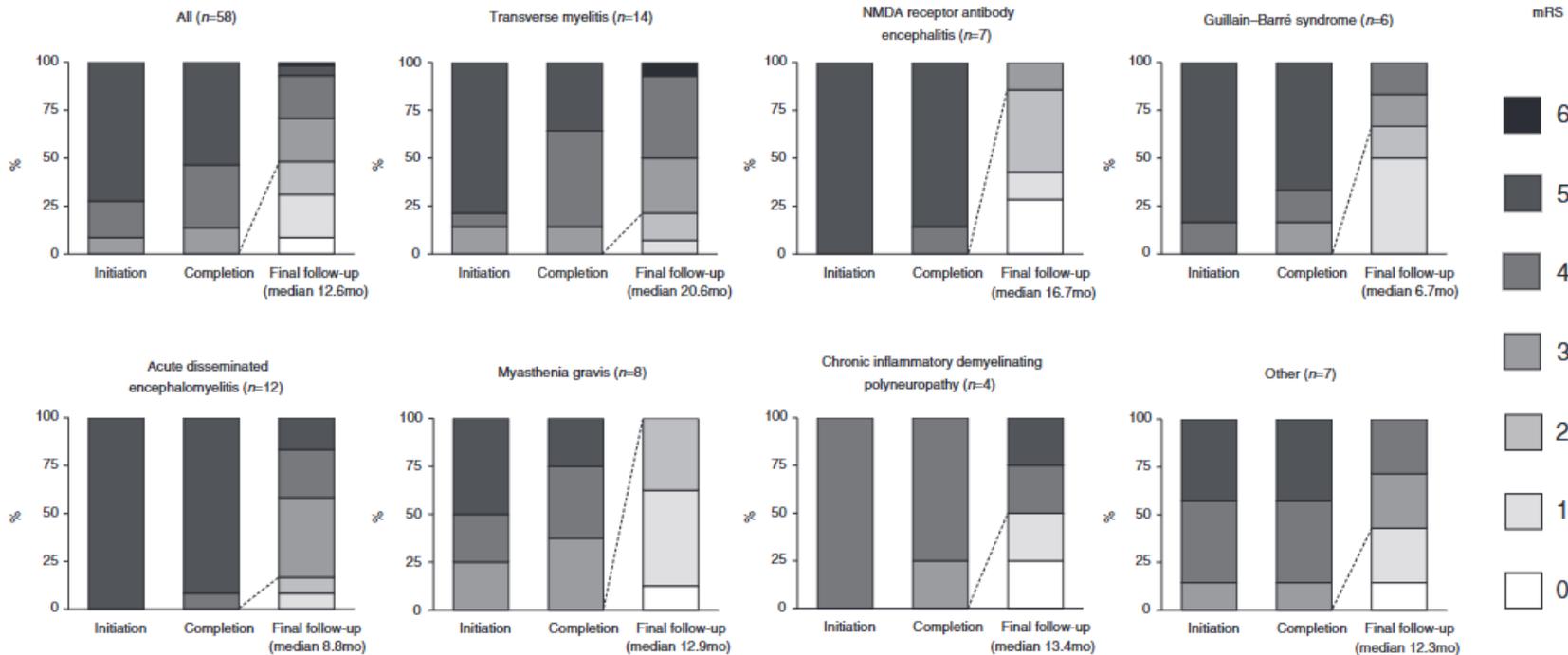
Inadequate response: consider repeat course of TPE, or other therapies as appropriate^d

4 centres UK

Mediane 6 PLEX (range 2–179)
durée médiane de 8 jours
(range 3–466).

Table 1: Characteristics of patients at initiation of plasma exchange

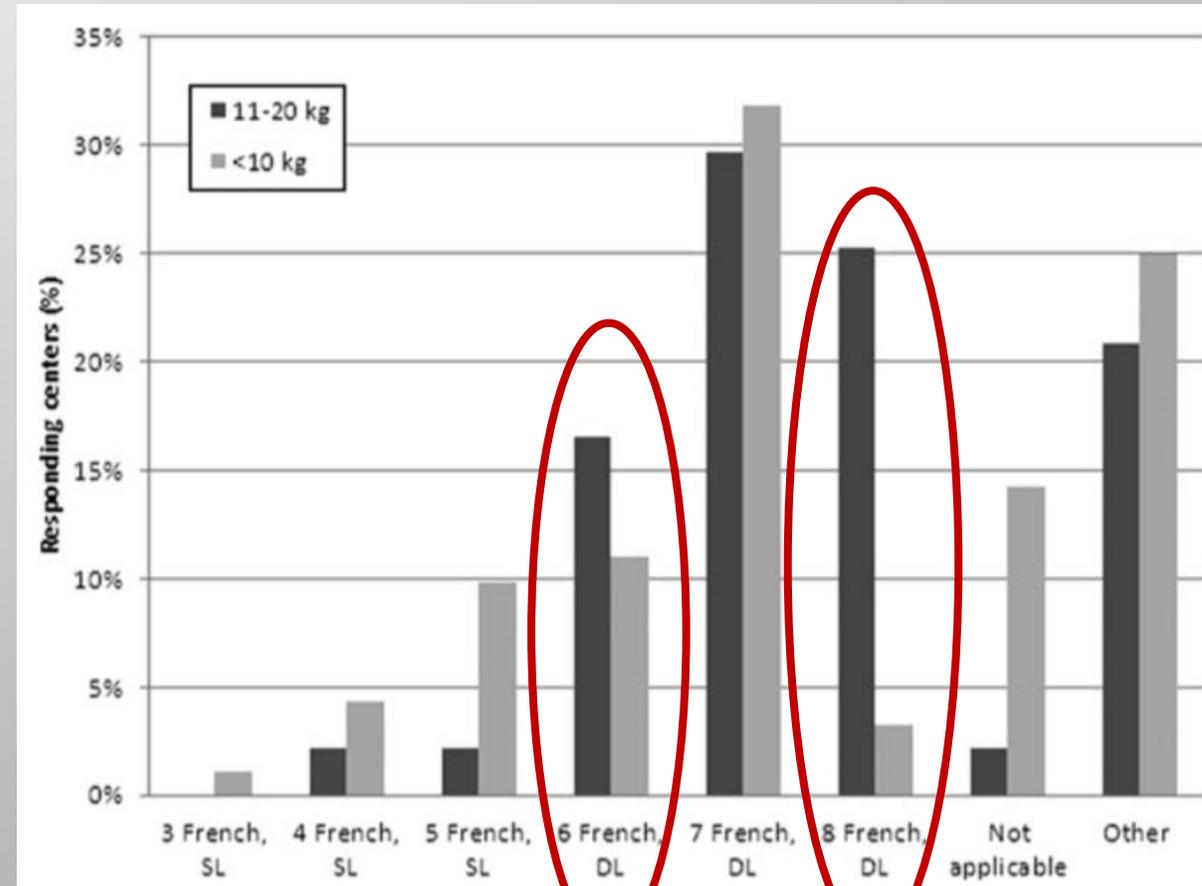
Indication for TPE	ASFA ^a category	n	Age in years, median (range)	Sex M/F	Duration of neurological illness at initiation of TPE in days, median (range)	Baseline neurological disability mRS, median (range)	PICU admission n (%)
All	–	58	9 (1–15)	25/33	25.5 (1–4575)	5 (3–5)	24/58 (41)
Transverse myelitis ^b	–	14	8 (1–13)	8/6	12 (3–83)	5 (3–5)	1/14 (7)
Acute disseminated encephalomyelitis	II	12	5.5 (1–14)	8/4	8.5 (1–66)	5 (5–5)	8/12 (67)
Myasthenia gravis	I	8	12 (9–15)	4/4	32 (11–248)	4.5 (3–5)	4/8 (50)
NMDAR-ab encephalitis	I	7	4 (2–13)	3/4	37 (24–92)	5 (5–5)	3/7 (43)
Guillain-Barré syndrome ^c	III	6	8 (1–15)	1/5	6 (3–49)	5 (5–5)	5/6 (83)
Chronic inflammatory demyelinating polyneuropathy	I	4	13 (3–14)	0/4	2100 (48–4575)	4 (4–4)	1/4 (25)
Presumed autoimmune encephalitis	II	4	14 (10–15)	0/4	46 (45–350)	4 (4–5)	0/4 (0)
Febrile infection-related epilepsy syndrome	–	2	5 (3–7)	1/1	16.5 (8–25)	5 (5–5)	2/2 (100)
Opsoclonus myoclonus	III	1	2	0/1	286	4	0/1 (0)



Eyre et al Develop med child
neuro. 2021

Particularités de la population pédiatrique

- Questionnaire international
- 90 centres apherèses thérapeutiques membres de l'AFSA
 - 40% des centres prennent en charge patients < 12 mois
 - 36% pas de limite de poids
 - Médiane poids limite 6,3Kgs
- **Le problème de la voie d'abord et du volume sanguin du circuit extra corporel...**



Problème du volume du circuit extra corporel en pédiatrie

- Volume sanguin en pédiatrie :
 - Prématuré: 100ml/kg
 - Nouveau-né et nourrisson < 3mois: 85 ml/kg
 - Nourrisson 3 mois et enfant: 75 ml/kg
 - Adolescent: 70 ml/kg (male) / 65 ml/kg (femme)
- Volume des circuits sanguins:

Prismax -Baxter®	Life 21 -Myltenii®®	Multifiltrate Fresenius®
TPE set 1000: 71ml - TPE set 2000: 125ml	Lignes = disque séparateur 63 ml	lignes Paed CRRT 54 ml MPS 2 Dry 180ml
Plasmafiltre TPE 2000: 41 ml Plasmafiltre TPE 1000: 23ml	Therasorb Ig OMNI 5 50 ml	PLASMAFLUX PSU 1S : 0.25 m2, 36 ml. MPS2 dry: 0.6 m2, 70 ml.

**Ne pas dépasser 10-15ml/kg
pour le circuit extra-
corporel)**

Complications des TPE

- 53 patients
- Avril 2006-oct 2022
- Age median 13ans
- 378 PLEX

- 58 patients
- Avril 2007-oct 2013
- Age median 9 ans
- 693 PLEX

Complication, n/N (%)	Complication severity*	
	All grades	Grades 3-4
Suspected line infection	11/67 (16%)	5/67 (8%)
Line obstruction	3/67 (5%)	0/67 (0%)
Line displacement	2/67 (3%)	0/67 (0%)
Excessive bleeding	1/67 (2%)	0/67 (0%)
Complications of anaesthetic	1/67 (2%)	1/67 (2%)

Type of Complication	<i>n</i> = 47	%
Nausea/vomiting	15	32
Drop in blood pressure	9	19
Pruritus	4	9
Headache	3	6
Non-infectious catheter-associated complications	20	43
Infectious catheter-associated complication	4	9
Sepsis	1	2
Other complications	19	40

Complication	Complication severity ^a	
	All grades, <i>n</i> =67 (%)	Grades 3-4, <i>n</i> =67 (%)
Hypocalcaemia	13 (19)	0 (0)
Anaemia	10 (15)	8 (12)
Hypotension	6 (9)	4 (6)
Coagulopathy	5 (8)	0 (0)
Hypokalaemia	3 (5)	0 (0)
Thrombocytopenia	3 (5)	1 (2)
Bradycardia	2 (3)	2 (3)
Hypertension	1 (2)	0 (0)
Hypophosphataemia	1 (2)	0 (0)
Hyperkalaemia	1 (2)	1 (2)
Rash	1 (2)	0 (0)

Avantages/ inconvénients EP vs IA en pédiatrie

EP:

Volume circuit extra corporel réduit (+/-)

Durée séance (+/-)

Risque réaction anaphylactique (Plasma)+++

Perte albumine, facteurs de coagulation=>
risque hémorragique

Immunologique:

Diminution des complexes immuns circulants

Sensibilisation des cellules productrices d'Ac aux
traitements immunosuppresseurs

Diminution des cytokines (IL6, TNF α) et
molécules d'adhésion (sICAM,sVCAM)

Modification Th1/Th2 des LT helpers

Prix séance TEP<IA



IA:

Efficacité sur épuration IgG (sauf IgG4)

+/- CIC

Pas de nécessité de plasma/albumine

Moins de complications allergiques

Tolérance hémodynamique +





Objectif :

- Evaluer à l'échelle nationale **l'efficacité et la sécurité des échanges plasmatiques (TEP) ou immunoadsorptions (IA)** chez les enfants présentant une pathologie neuroinflammatoire

Etude rétrospective multicentrique nationale

12 centres

154 patients ayant bénéficié TEP ou IA pour une pathologie neuro-inflammatoire

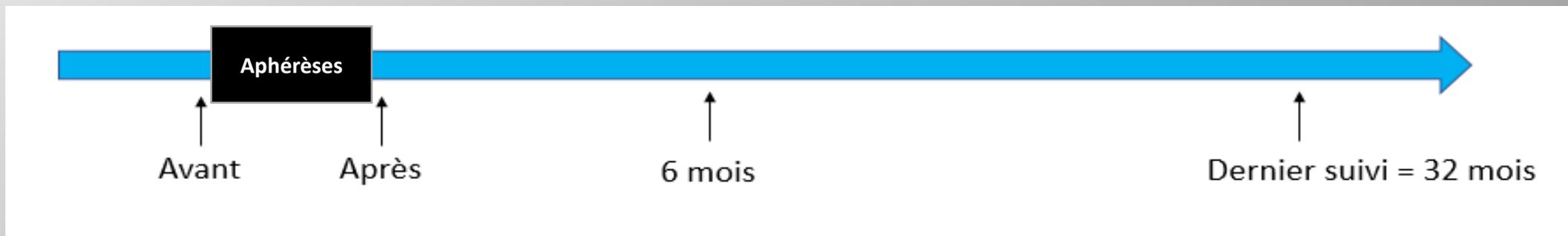
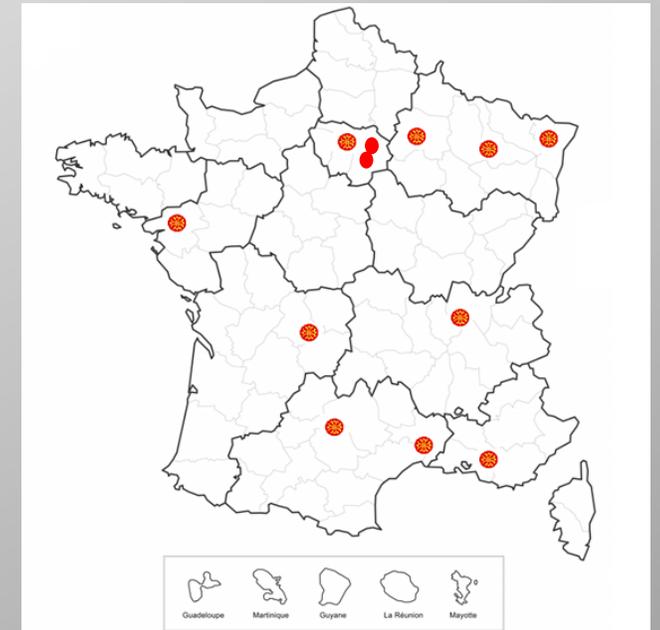
1037 TEP / 495 IA

Critère de jugement principal : amélioration du score de Rankin modifié (mRS) en début , fin de TPE/IA à M6 et à long terme

Critères de jugements secondaires :

Comparaison IA/TEP

effets indésirables TEP/IA



Take Home Message

- ❖ TPE et IA sont utiles dans les pathologies neuroinflammatoires pédiatriques
- ❖ Intérêt en première intention dans les formes sévères en association d'autres thérapies (SGB, Encéphalites r-NMDA, NMOSD)
- ❖ **Bonne tolérance des TPE/IA avec faible taux de complications graves**
- ❖ **Nécessité équipe IDE/puericultrices formés. Coopération multidisciplinaire « aphérèse »/neurologue pédiatre**
- ❖ Pour l'avenir:
 - ❖ Elaboration de protocoles communs indications TPE/IA en neurologie pédiatrique
 - ❖ Essais contrôlés randomisés pédiatriques (MOGAD....)

Merci pour votre attention