

2 & 3 décembre 2016

Bordeaux Hôtel Pullman



17^{èmes} Journées
d'Urgences Pédiatriques du Sud-Ouest



Münchhausen par procuration: les difficultés du clinicien

Leïla LAZARO

Centre Hospitalier de la Côte Basque



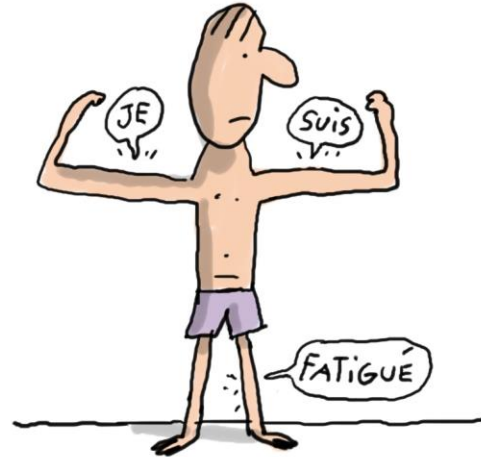
Histoires vraies



Histoires vraies



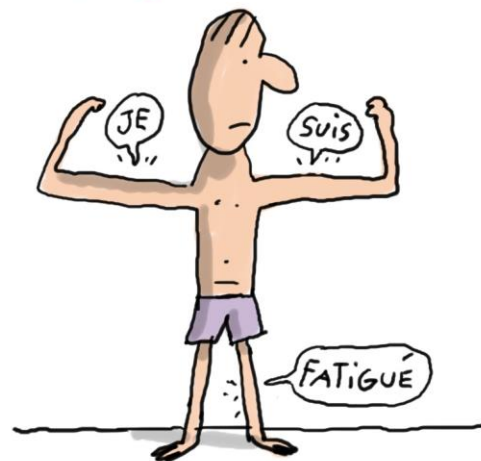
 MALADIE NEUROMUSCULAIRE



Histoires vraies



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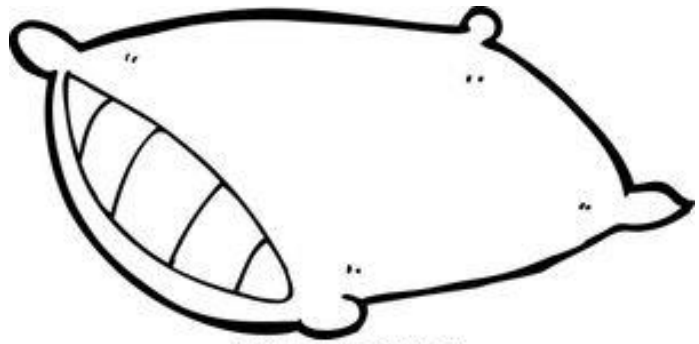
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Histoires vraies



Histoires vraies



Un peu d'histoire



Baron Karl Friedrich Hieronymus Von Munchhausen

Un peu d'histoire



Richard ASHER 1951
The Lancet

JUPSO 2016

THE LANCET

MUNCHAUSEN'S SYNDROME

[Feb. 10, 1951] 339

Special Articles

MUNCHAUSEN'S SYNDROME

RICHARD ASHER
M.D. Lond., M.R.C.P.

HERE is described a common syndrome which most doctors have seen, but about which little has been written. Like the famous Baron von Munchausen, the persons affected have always travelled widely; and their stories, like those attributed to him,¹ are both dramatic and untruthful. Accordingly the syndrome is respectfully dedicated to the baron, and named after him.

The patient showing the syndrome is admitted to hospital with apparent acute illness supported by a plausible and dramatic history. Usually his story is largely made up of falsehoods; he is found to have attended, and deceived, an astounding number of other hospitals; and he nearly always discharges himself against advice, after quarrelling violently with both doctors and nurses. A large number of abdominal scars is particularly characteristic of this condition.

That is a general outline; and few doctors can boast that they have never been hoodwinked by the condition. Often the diagnosis is made by a passing doctor or sister, who, recognising the patient and his performance, exclaims: "I know that man. We had him in St. Quindine's two years ago and thought he had a perforated ulcer. He's the man who always collapses on buses and tells a story about being an ex-submarine commander who was tortured by the Gestapo." Equally often, the trickster is first revealed in the hospital dining-room, when, with a burst of laughter, one of the other residents exclaims: "Good heavens, you haven't got Luella Priskins in again, surely? Why she's been in here three times before and in Barts, Mary's, and Guy's as well. She sometimes comes in with a different name, but always says she's coughed up pints of blood and tells a story about being an ex-opera-singer and helping in the French resistance movement."

DIAGNOSIS

It is almost impossible to be certain of the diagnosis at first, and it requires a bold casualty officer to refuse admission. Usually the patient seems seriously ill and is admitted unless someone who has seen him before is there to expose his past. Experienced front-gate porters are often invaluable at doing this.

The following are useful pointers:

1. (Already mentioned) a multiplicity of scars, often abdominal.
2. A mixture of treachery and evasiveness in manner.
3. An immediate history which is always acute and harrowing yet not entirely convincing—overwhelmingly severe abdominal pain of uncertain type, metastasising blood-loss unsupported by corresponding pallor, dramatic loss of consciousness, and so forth.
4. A wallet or handbag stuffed with hospital attendance cards, insurance claim forms, and litigious correspondence.

If the patient is not recognised by an old acquaintance, the diagnosis is only gradually revealed by inquiries at other hospitals. Some have given so much trouble elsewhere they have been placed on hospital black-lists. Often the police are found to know the patient and can give many helpful details. Gradually the true history is pieced together and the patient's own story is seen to be a matrix of fantasy and falsehood, in which fragments of complete truth are surprisingly imbedded. Just as the patient's story is not wholly false, so neither are all the symptoms; and it must be recognised that

these patients are often quite ill, although their illness is shrouded by duplicity and distortion. When the whole truth is known, past history sometimes reveals drug-addiction, mental-hospital treatment, or prison sentences, but these factors are not constant, and the past may consist solely in innumerable admissions to hospitals and evidence of pathological lying. Often a real organic lesion from the past has left some genuine physical signs which the patient uses (to quote Pook Bah)² "to give artistic verisimilitude to an otherwise bald and unconvincing narrative."

SOME CHARACTERISTIC FEATURES

Most cases resemble organic emergencies. Well-known varieties are:

1. The acute abdominal type (laparotomophilia migrans), which is the most common. Some of these patients have been operated on so often that the development of genuine intestinal obstruction from adhesions may confine the picture.
2. The haemorrhagic type, who specialise in bleeding from lungs or stomach, or other blood-loss. They are colloquially known as "haemoptysis merchants" and "haematemesis merchants."
3. The neurological type, presenting with paroxysmal headache, loss of consciousness, or peculiar fits.

The most remarkable feature of the syndrome is the apparent senselessness of it. Unlike the malingerer, who may gain a definite end, these patients often seem to gain nothing except the discomfort of unnecessary investigations or operations. Their initial tolerance to the more brutish hospital measures is remarkable, yet they commonly discharge themselves after a few days with operation wounds scarcely healed, or intravenous drips still running.

Another feature is their intense desire to deceive everybody as much as possible. Many of their falsehoods seem to have little point. They lie for the sake of lying. They give false addresses, false names, and false occupations merely from a love of falsehood. Their effrontery is sometimes formidable, and they may appear many times at the same hospital, hoping to meet a new doctor upon whom to practise their deception.

POSSIBLE MOTIVES

Sometimes the motive is never clearly ascertained, but there are indications that one of the following mechanisms may be involved:

1. A desire to be the centre of interest and attention. They may be suffering in fact from the Walter Mitty syndrome,³ but instead of playing the dramatic part of the surgeon, they submit to the equally dramatic rôle of the patient.
2. A grudge against doctors and hospitals, which is satisfied by frustrating or deceiving them.
3. A desire for drugs.
4. A desire to escape from the police. (These patients often swallow foreign bodies, interfere with their wounds, or manipulate their thermometers.)
5. A desire to get free board and lodgings for the night, despite the risk of investigations and treatment.

Supplementing these strange motives, there probably exists some strange twist of personality. Perhaps most cases are hysterics, schizophrénics, maniacs, or psychopaths of some kind; but as a group they show such a constant pattern of behaviour that it is worth considering them together.

ILLUSTRATIVE CASE-RECORDS

Three cases of the abdominal type of Munchausen's syndrome are described below; for they show clearly the typical features of the advanced form of the disease. Many other milder forms have been encountered, but it would be tedious if more were described. All the

¹ Rappe, R. E., et al. (1785) *Stougar Travels, Campaigns and Adventures of Baron Munchausen*. London: Cresset Press, 1948.

² Thurber, J. *The Secret Life of Walter Mitty*. My World and Welcome to It. London, 1942.

Un peu d'histoire

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Hospital Practice

MUNCHHAUSEN SYNDROME BY PROXY THE HINTERLAND OF CHILD ABUSE

ROY MEADOW

Department of Paediatrics and Child Health,
Sacroft Hospital, Leeds

Summary Some patients consistently produce false stories and fabricate evidence, so causing themselves needless hospital investigations and operations. Here are described parents who, by falsification, caused their children innumerable harmful hospital procedures—a sort of Munchausen syndrome by proxy.

Introduction

Doctors dealing with young children rely on the parents' recollection of the history. The doctor accepts that history, albeit sometimes with a pinch of salt, and it forms the cornerstone of subsequent investigation and management of the child.

A case is reported in which over a period of six years, the parents systematically provided fictitious information about their child's symptoms, tampered with the urine specimens to produce false results and interfered with hospital observations. This caused the girl innumerable investigations and anaesthetic, surgical, and radiological procedures in three different centres.

The case is compared with another child who was intermittently given toxic doses of salt which again led to massive investigation in three different centres, and ended in death. The behaviour of the parents of these two cases was similar in many ways. Although in each case the end result for the child was "non-accidental injury", the long-running saga of hospital care was reminiscent of the Munchausen syndrome, in these cases by proxy.

Case-reports

FIRST CASE

Kay was referred to the paediatric nephrology clinic in Leeds at the age of 6 because of recurrent illnesses in which she passed foul-smelling, bloody urine. She had been investigated in two other centres without the cause being found.

In the child's infancy, her mother had noticed yellow pus on the nappies, and their doctor had first prescribed antibiotics for suspected urine infection when Kay was 8 months old. Since then, she had had periodic courses of antibiotics for presumed urine infection. Since the age of 3 she had been on continuous antibiotics which included co-trimoxazole, amoxicillin, nalidixic acid, nitrofurantoin, ampicillin, gentamicin, and ticarcillin. These treatments had themselves caused drug rashes, fever, and candidiasis, and she had continued to have intermittent bouts of lower abdominal pain associated with fever and foul-smelling, infected urine often containing frank blood. There was intermittent vulval soreness and discharge.

The parents were in their late 30s. Father who worked mainly in the evenings and at night, was healthy. The mother had had urinary-tract infections. The 3-year-old brother was healthy.

At the time of referral, she had already been investigated at a district general hospital and at a regional teaching hospital.

Investigations had included two urograms, micturating cystourethrograms, two gynaecological examinations under anaesthetic, and two cystoscopies. The symptoms were unexplained and continued unabated. She was being given steadily more toxic chemotherapy. Bouts were recurring more often and everyone was mystified by the intermittent nature of her complaint and the way in which purulent, bloody urine specimens were followed by completely clear ones a few hours later. Similarly, foul discharges were apparent on her vulva at one moment, but later on the same day her vulva was normal.

On examination she was a healthy girl who was growing normally. The urine was bloodstained and foul. It was strongly positive for blood and albumin and contained a great many leucocytes and epithelial cells. It was heavily infected with *Escherichia coli*.

The findings strongly suggested an ectopic ureter or an infected cyst draining into the urethra or vagina. Yet previous investigations had not disclosed this. Ectopic ureters are notoriously difficult to detect, and, after consultation with colleagues at the combined paediatric/urology clinic, it was decided to investigate her immediately she began to pass foul urine. No sooner was she admitted than the foul discharge stopped before cystoscopy could be done. More efficient arrangements were made for the urological surgery concerned to be contacted immediately she should arrive in Leeds, passing foul urine. This was done three times (including a bank holiday and a Sunday). No source of the discharge was found. On every occasion it cleared up fast. Efforts to localise the source included further radiology, vaginogram, urethrogram, barium enema, suprapubic aspiration, bladder catheterisation, urine cultures, and exfoliative cytology. During these investigations, the parents were most cooperative and Kay's mother always stayed in hospital with her (mainly because they lived a long way away). She was concerned and loving in her relationship with Kay, and yet sometimes not quite so worried about the possible cause of the illness as were the doctors. Many of the crises involved immediate admission and urgent anaesthetics for examinations or cystoscopy, and these tended to occur most at weekend holiday periods. On one bank holiday, five consultants came into the hospital specifically to see her.

The problem seemed insoluble and many of the facts did not make sense. The urinary pathogens came and went at a few minutes' notice; there would be one variety of *E. coli* early in the morning and then after a few normal specimens, an entirely different organism such as *Proteus* or *Streptococcus faecalis* in the evening. Moreover, there was something about the mother's temperament and behaviour that was reminiscent of the mother described in case 2, so we decided to work on the assumption that everything about the history and investigations were false. Close questioning revealed that most of the abnormal specimens were ones that at some stage or other had been left unsupervised in the mother's presence.

This theory was tested when Kay was admitted with her mother and all urine specimens were collected under strict supervision by a trained nurse who was told not to let the urine out of her sight from the moment it passed from Kay's urethra to it being tested on the ward by a doctor and then delivered to the laboratory. On the fourth day, supervision was deliberately relaxed slightly so that one or two specimens were either left for the mother to collect or collected by the nurse and then left in the mother's presence for a minute before being taken away. On the first 3 days, no urine specimen was abnormal. On the first occasion that the mother was left to collect the specimen (having been instructed exactly how to do so), she brought a heavily bloodstained specimen containing much debris and bacteria. A subsequent specimen collected by the nurse, was completely normal. This happened on many occasions during the next few days. During a 7-day period, Kay emptied her bladder 57 times; 45 specimens were normal, all of these being collected and supervised by a nurse; 12 were grossly abnormal, containing blood and different organisms, all these having been collected by the mother or left in her presence.



Roy MEADOW 1977
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Un peu d'histoire



Richard ASHER 1951



Roy MEADOW 1977

ROSENBERG ...

LIBOW et SCHREIER ...

Définition

- Troubles factices dans le **DSM V**
- Abus à enfant dans la **CIM-10**
- **Asher 1951**: « Situation dans laquelle une personne fabrique de façon persistante des signes à son enfant, lui causant des examens physiques et des traitements innombrables, douloureux et inutiles, dont l'évolution peut être mortelle »
- **Libow et Schreier** définissent 3 formes
 - Demandeur d'aide ou *Help Seekers*:
 - Inducteurs actifs ou *Active inducers*
 - Dépendants aux médecins ou *Doctors addicts*

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 - **Demandeur d'aide ou *Help Seekers***: mères déprimées ou anxieuses qui falsifient ou provoquent des symptômes organiques chez leur enfant dans le but de le faire hospitaliser et d'être ainsi rassurées par les pédiatres. Ces parents ne dénie pas leurs conduites de falsification et reconnaissent le besoin d'être aidés dans leurs responsabilités parentales. Ils acceptent souvent avec soulagement l'aide proposée

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- **Libow et Schreier** définissent 3 formes
 - **Inducteurs actifs ou *Active inducers***: ce sont les cas les plus fréquemment rapportés dans la littérature, caractérisés par des symptômes graves activement induits par un parent qui agit directement sur le corps de l'enfant comme lors des intoxications ou des suffocations. Les enfants sont souvent très jeunes en âge pré-scolaire. Dans ce cas, le parent nie toute responsabilité dans la symptomatologie.

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 - **Dépendants aux médecins ou *Doctors addicts***: il s'agit de mères obsédées par la poursuite d'un diagnostic et l'application des traitements. Convaincues de la réalité de la maladie de leur enfant, elles peuvent installer une attitude suspicieuse, revendicative vis-à-vis des équipes médicales voire un comportement paranoïaque. Les enfants sont souvent plus âgés et atteints de maladies psychosomatiques.

Epidémiologie

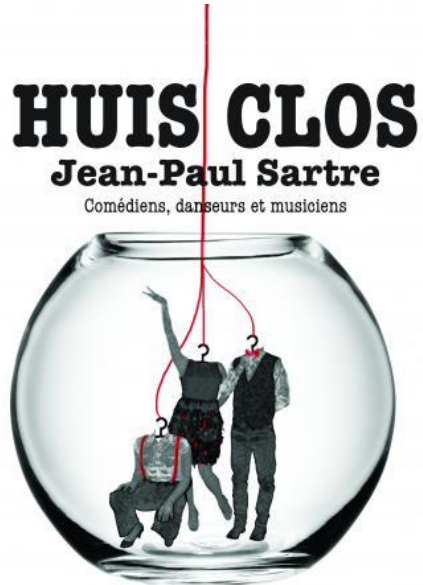
- Forme de maltraitance longtemps méconnue
- Incidence méconnue en France
- Garçon = Fille
- Age moyen au diagnostic = 20 à 40 mois
- 6 à 10% de mortalité
- Mère responsable dans 85% des cas
- Morbidité pour $\frac{1}{4}$ liée aux actes médicaux et pour $\frac{3}{4}$ à l'association des actes médicaux + actes du parent agresseur



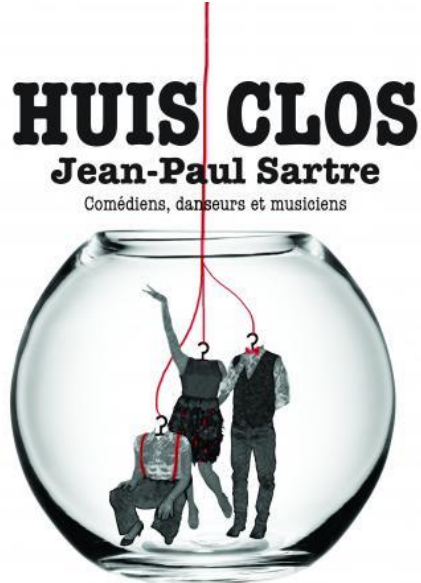
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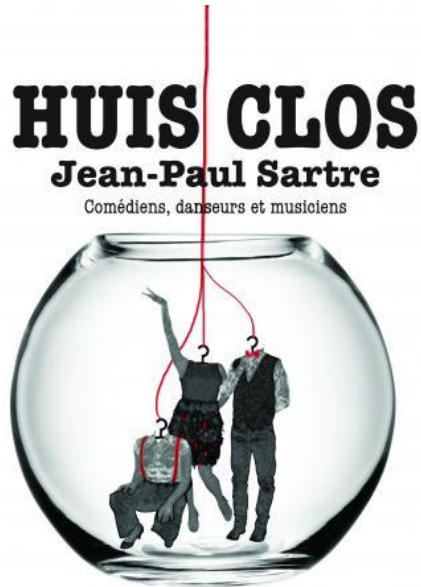
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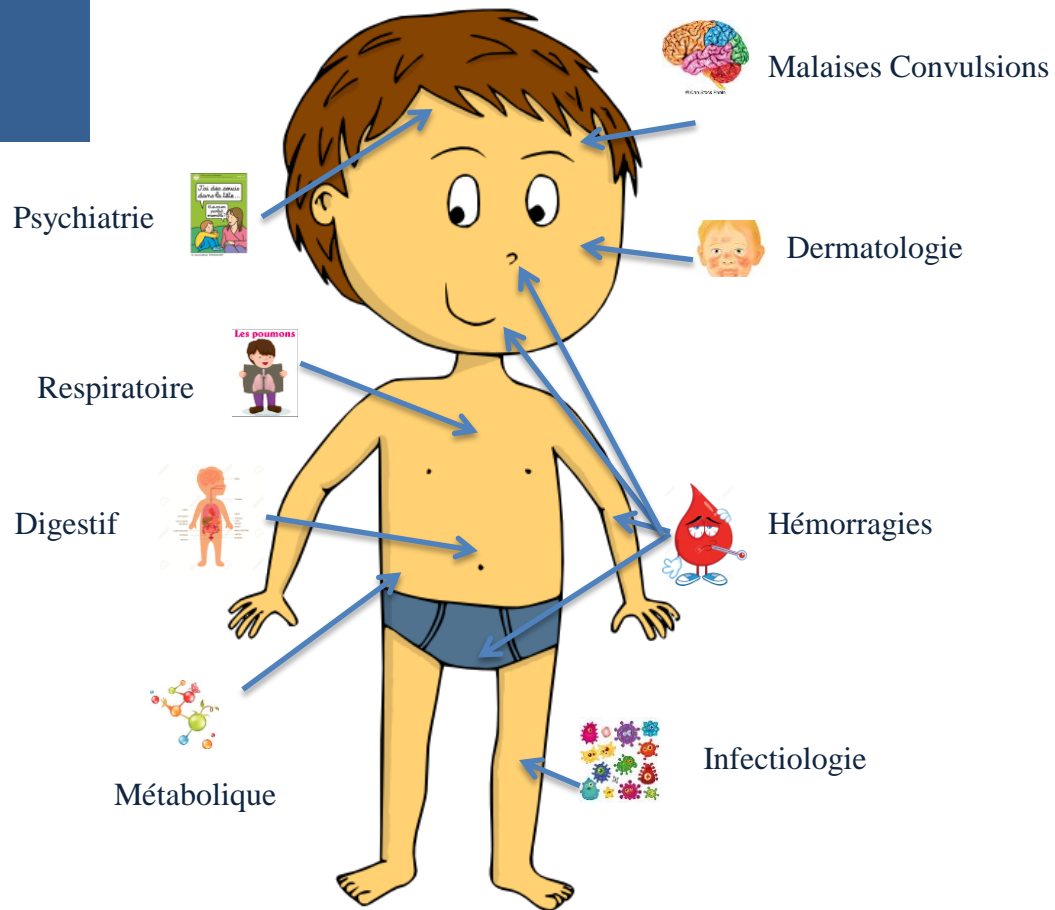
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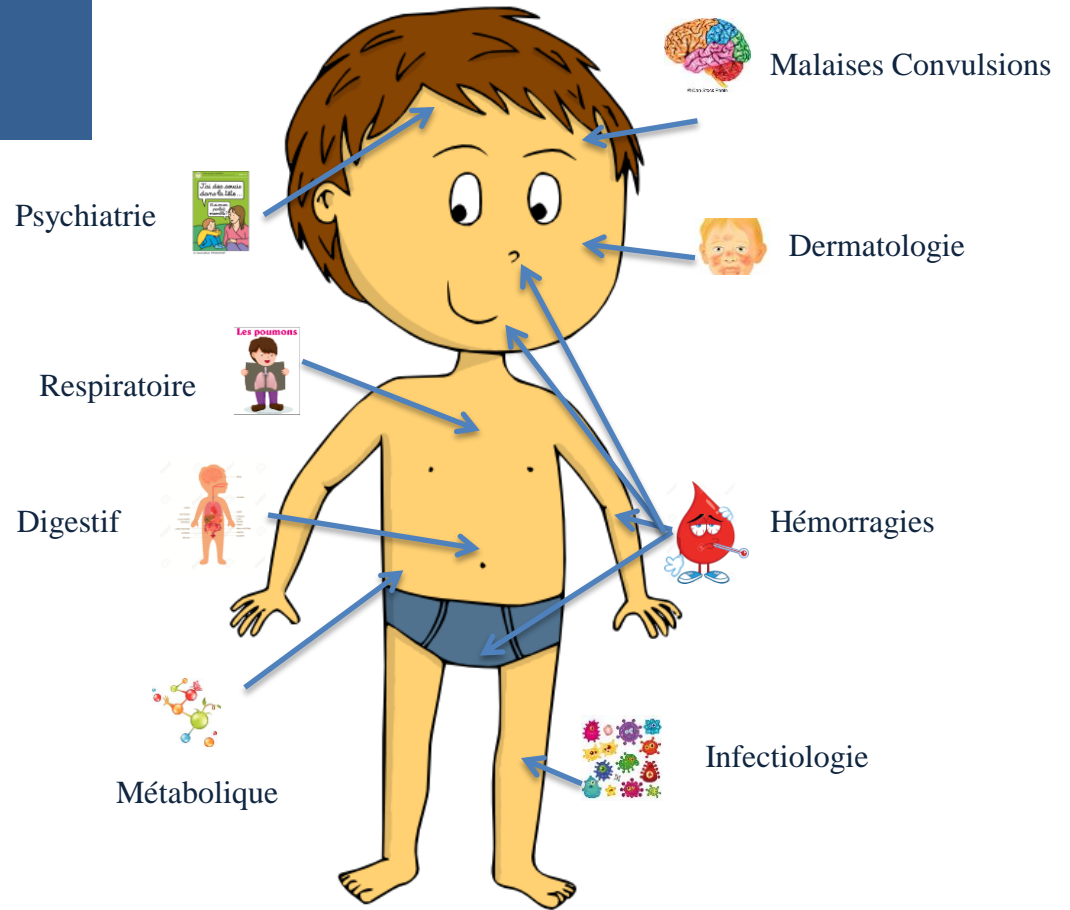


La clinique



La clinique

- ⇒ Symptômes variables multiples insolites
- ⇒ Substrat organique parfois sous-jacent
- ⇒ Symptômes psychologiques secondaires au SMPP
- ⇒ Formes atypiques en psychiatrie



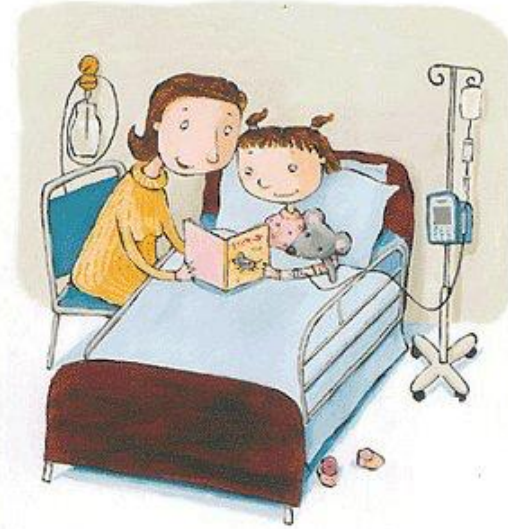
Signes d'Alerte de Meadow

- Maladie prolongée inexplicable
- Signes incongrus et inhabituels
- N'apparaissant qu'en présence de la mère
- Disparaissant en son absence
- Traitements inefficaces et mal tolérés
- Allergies multiples et variées
- Mère très présente au chevet de son enfant
- Très concernée par la démarche médicale
- Et paradoxalement peu inquiète des souffrances de son enfant
- Père absent dans les démarches médicales
- Maladies rares
- Décès dans la fratrie



Qui est cette mère ?

- Femme intelligente, mariée
- Relations sociales pauvres
- Vie conjugale peu satisfaisante
- Mère admirable, dévouée
- Relation privilégiée avec le monde médical
- (Appartenant au monde médical ou de la petite enfance)
- Souvent ATCD de maltraitance
- ATCD de maladie somatique dans la petite enfance
- Pathologie psychiatrique rare
- Tous les milieux sociaux
- Aggravation de la sévérité et du risque de décès en lien avec les inégalités socio-économiques



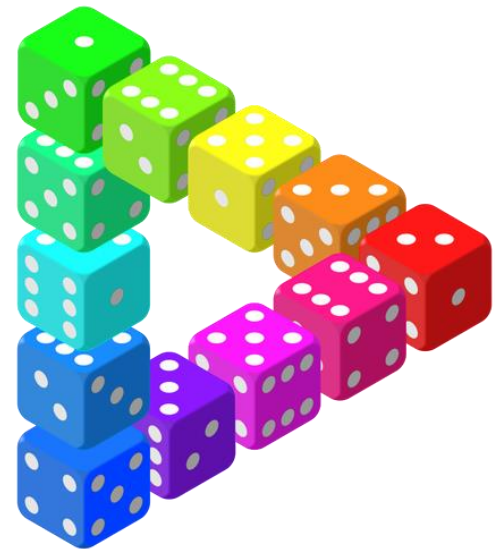
Qui est cette mère ?



- Par procuration, elles devient le centre de toutes les attentions
- Relation symbiotique à son enfant
- N'autorise pas à son enfant une quelconque forme d'autonomie
- Relation ambivalente de dépendance et d'hostilité avec les médecins
- Semble anesthésiée affectivement

Qu'est-ce que le SMPP ?

- *Ce n'est pas une pathologie psychiatrique*
- *C'est:*
 - Un Trouble grave de la relation Parent-Enfant
 - Un Trouble grave de la relation Parent-Médecin
 - Une Triangulaire Parent-Enfant-Médecin



Autres formes cliniques

- Munchhäusen *avec* procuration Adulte – Adulte
- Munchhäusen *sans* procuration

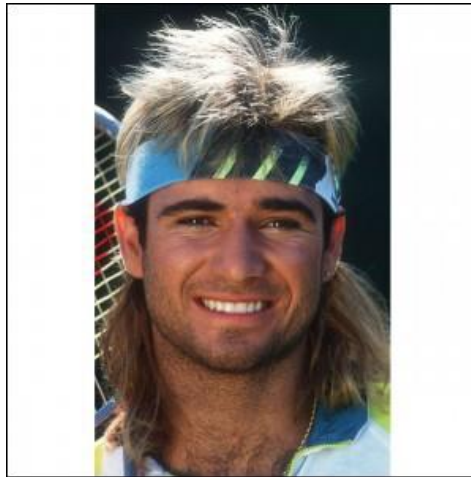


Autres formes cliniques

Syndrome de Réussite par Procuration

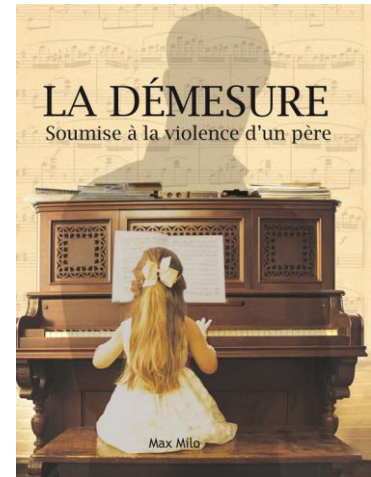


Mozart



André Agassi

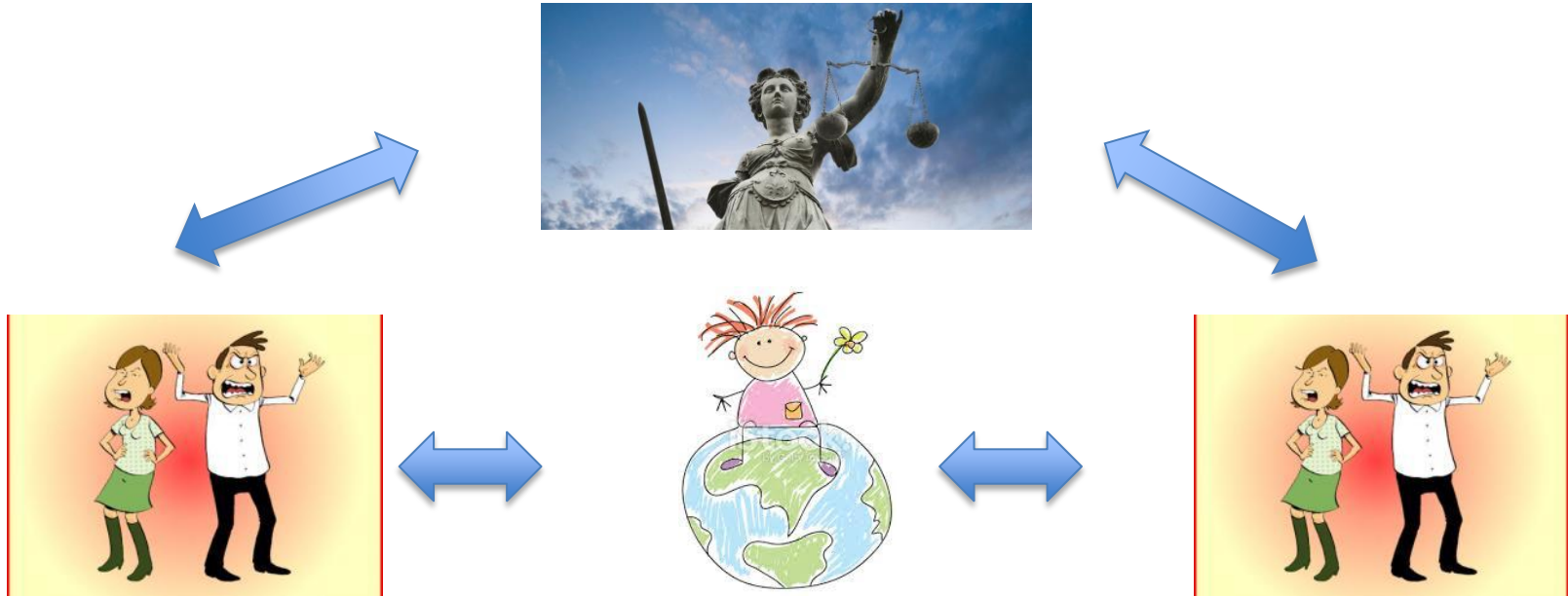
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Céline Raphaël

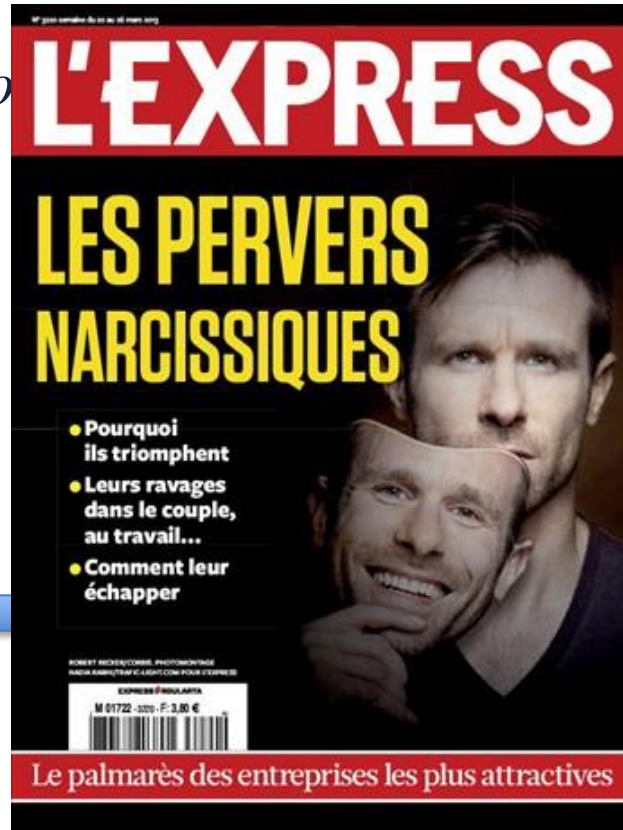
Autres formes cliniques

Syndrome d'Aliénation Parentale



Autres formes cliniques

Syndrome d'Aliénatio



Place du médecin

- ① Vis-à-vis de l'enfant et de la famille
- ① Vis-à-vis du monde médical
- ① Vis-à-vis de la justice

Place du médecin

Vis-à-vis de l'enfant et de la famille



- ① Garder une alliance thérapeutique
- ② Garder des échantillons biologiques à visée médico-légale
- ③ Arrêter ou limiter les investigations médicales
- ④ Isoler l'enfant du parent pour observation
- ⑤ Protéger l'enfant

Place du médecin

Cela suppose d'accepter en tant que médecin:

- d'avoir été instrumentalisé par le parent acteur
- d'avoir été détourné de sa fonction de soignant
- de s'être trompé dans le diagnostic
- de ne pas être dans l'affect

=> *Atteinte narcissique* pour le médecin

=> *Problème de connaissance* du SMPP

Place du médecin



Vis-à-vis du monde médical

- ① Contacter tous les professionnels autour de l'enfant
- ② Retracer le parcours médical de l'enfant (Assurance maladie)
- ③ Définir un référent médical
- ④ Portage de l'équipe soignante pour éviter les partis pris
- ⑤ Coordonner la prise en charge

Place du médecin

Cela suppose pour le corps médical:

- de se mettre d'accord sur le diagnostic
- de légitimer un des médecins comme référent
- de ne plus répondre aux sollicitations de la famille
- d'être persévérant dans la démarche médicale
- de travailler en partenariat avec le monde judiciaire

Place du médecin

Vis-à-vis de la justice



- ① Echange préalable téléphonique avec procureur ou substitut
- ② Signalement judiciaire (plutôt que IP)
- ③ Intérêt d'une expertise psychiatrique du parent à discuter
- ④ Problème du niveau de preuve à apporter ?

Place du médecin

Cela suppose pour le monde judiciaire:

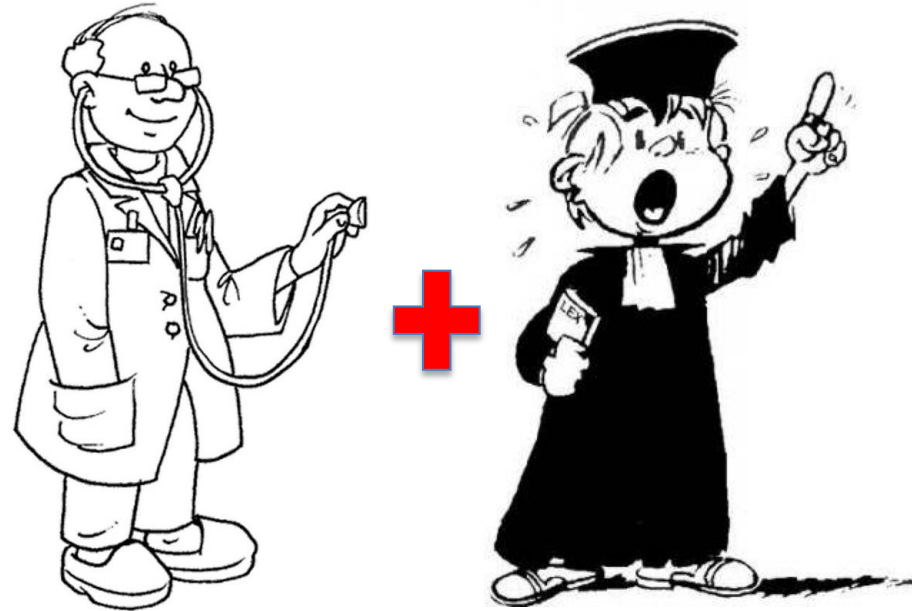
- d'être formé au SMPP
- que les expertises soient réalisées par des professionnels au fait du SMPP
- de travailler en partenariat avec le monde médical

Première Conclusion: Le Partenariat Médecine - Justice

Le diagnostic n'est définitivement certain qu'à la disparition des symptômes lorsque l'enfant n'est plus en présence du parent acteur (attention aux troubles du comportement secondaires du SMPP)

=>Le monde médical évoque le diagnostic

=>Le monde judiciaire donne les moyens de confirmer le diagnostic en notifiant la séparation de l'enfant de sa famille



Prévention du SMPP

- *Situations à risque:*
 - Prématurité
 - Maternité
 - Handicap
 - Maladie chronique somatique ou psychiatrique
- *Evolution à l'âge adulte méconnue*
- *Risque de Reproduction trans-générationnelle du SMPP*

Seconde Conclusion: La Prévention

- *Primaire*
 - ⇒ Reconnaissance des situations à risque en périnatalité
- *Secondaire*
 - ⇒ Extraire l'enfant du milieu familial et le suivre jusqu'à l'âge adulte
- *Tertiaire*
 - ⇒ Prévention des violences intra-familiales
 - ⇒ Thérapies de 3^e génération adaptées au stress post-traumatique (retentissement sur le développement précoce cérébral)

2 & 3 décembre 2016

Bordeaux Hôtel Pullman



17^{èmes} Journées
d'Urgences Pédiatriques du Sud-Ouest



Merci