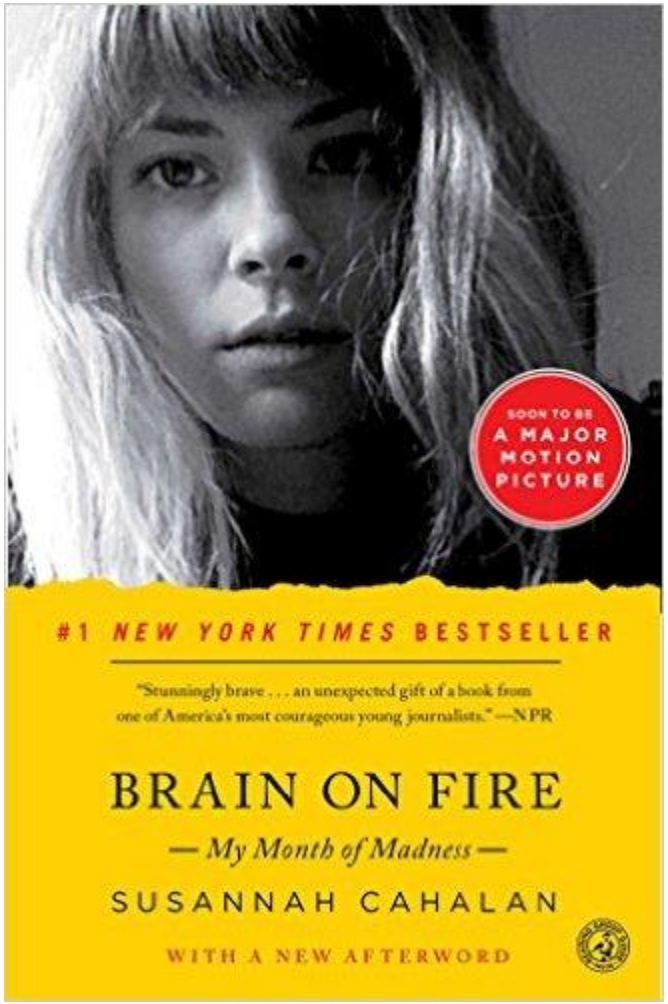
The Anti-NMDA Receptor Encephalitis

Anti-NMDAR Encephalitis
抗NMDA受體腦炎

血清組 李哲睿

UCL

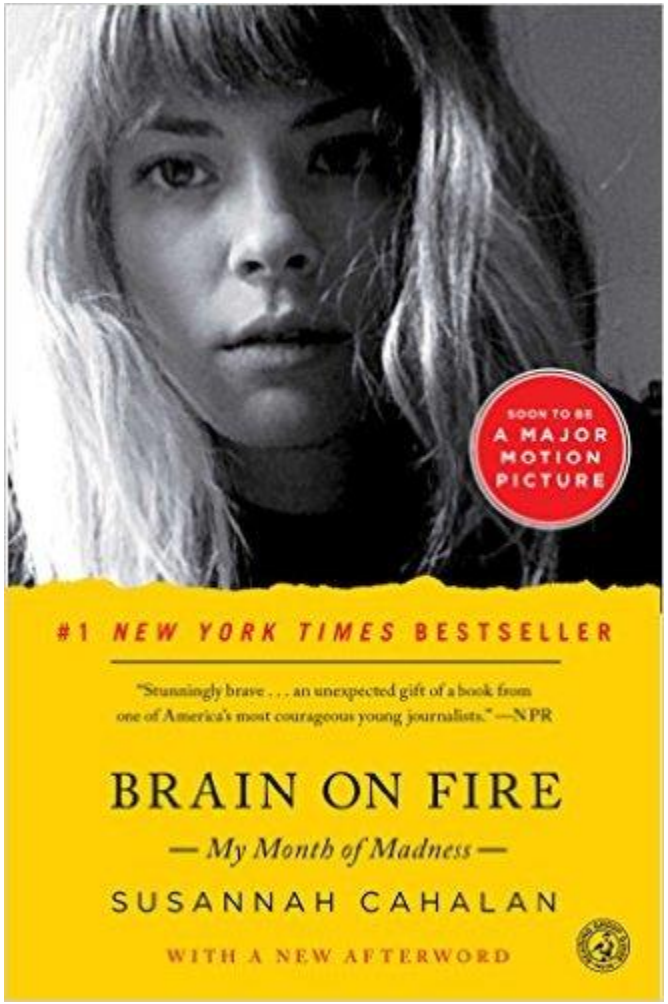
Overview



- **Anti-NMDAR Encephalitis**
  (抗NMDA受體腦炎)
  - Paraneoplastic encephalitis
    (附腫瘤神經性腦炎)
  - Immune-mediated encephalitis
    (免疫性腦炎)
  - Encephalopathic autoimmune disease
    (腦炎性自體免疫性疾病)

抗NMDA受體腦炎是一種急性腦炎，症狀嚴重是有可能會致命

Overview



- **Anti-NMDAR Encephalitis**
- (抗NMDA受體腦炎)

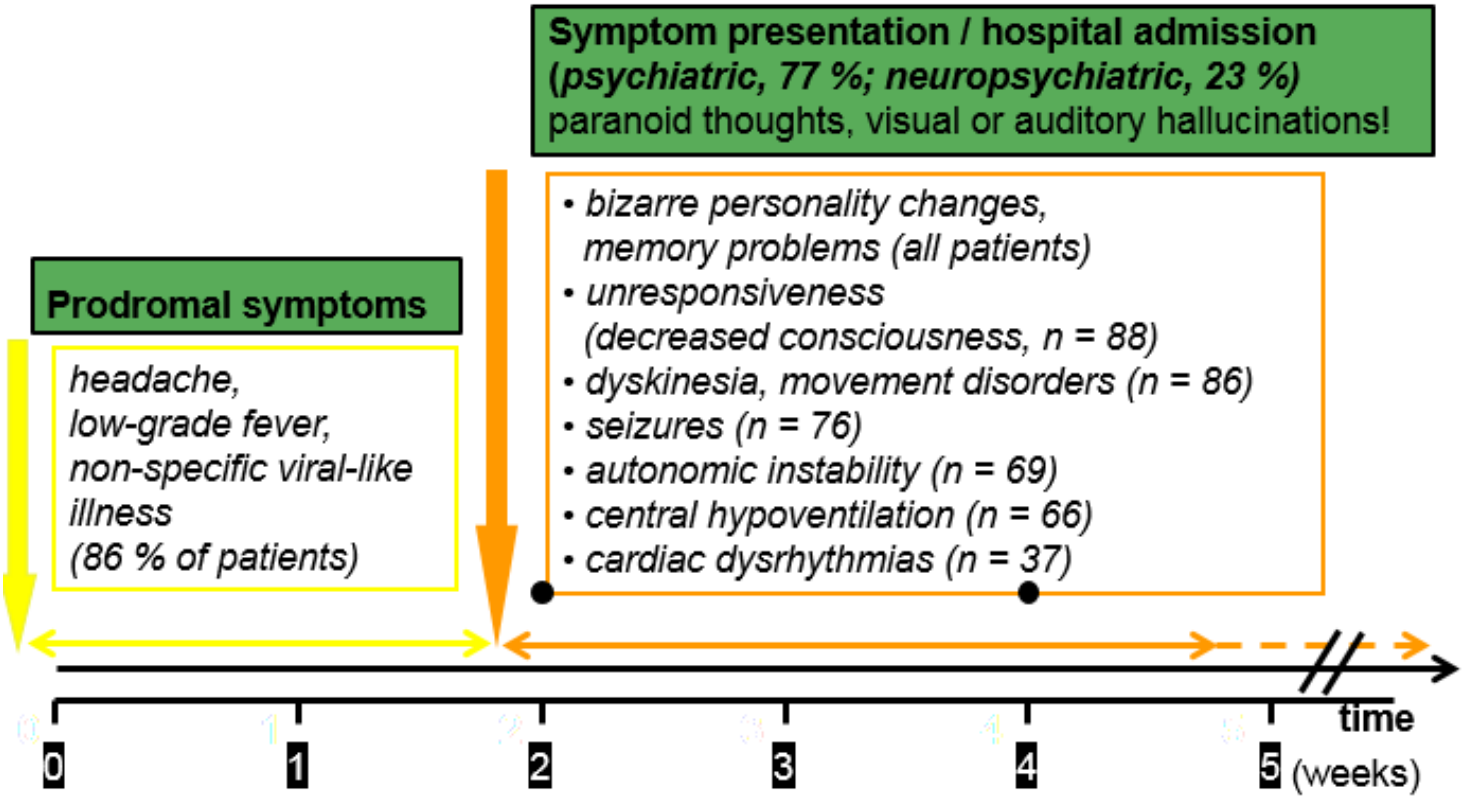
- 起初於2007年被報導出來，有廣泛症狀被診斷為該疾病

- 好發於年輕女性 (20 - 50 歲)

- 早期病毒性感染症狀盛行
  症狀有點像是病毒性感冒
- 中期異常發生精神性疾病等症狀

- 疾病的發生可能伴隨著卵巢畸胎瘤

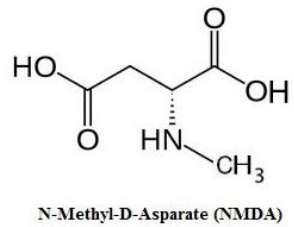
臨床症狀



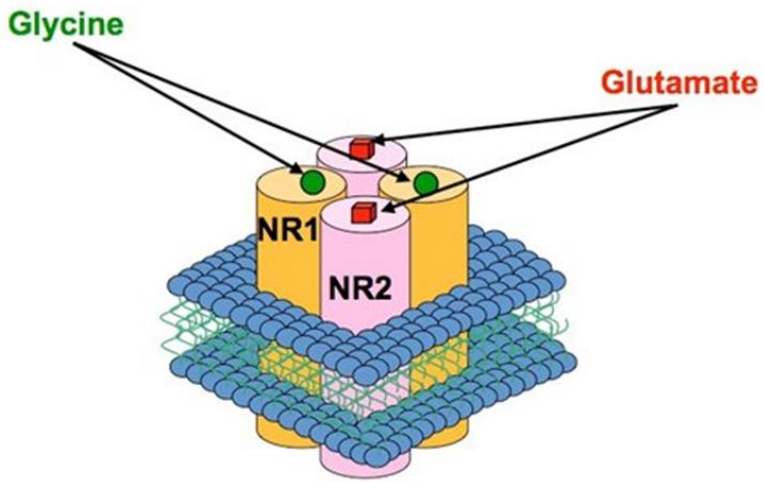
- Psychiatric (精神性疾病)
- Seizures (癲癇)
- Decrease level of consciousness (意識水平低下)
- Autonomic instability (自律神經失調)
- Central hypoventilation (中樞性換氣不足症候群)

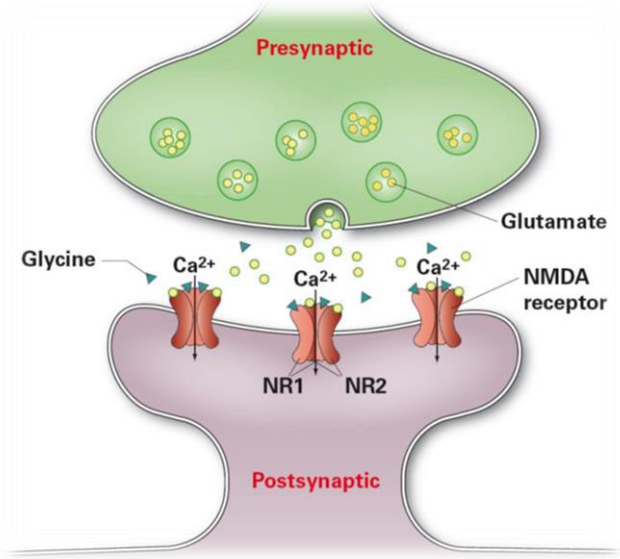
Mechanism and Pathophysiology

- NMDAR
  - N-methyl-D-aspartate receptor (N-甲基-D-天冬氨酸)
  - NR1(bind glycine)以及NR2 (bind glutamate)的異構物
  - 突觸的訊息傳遞, 神經細胞的重塑, 樹突的拓展, 海馬迴長期記憶區的調控, 記憶的生成及學習能力

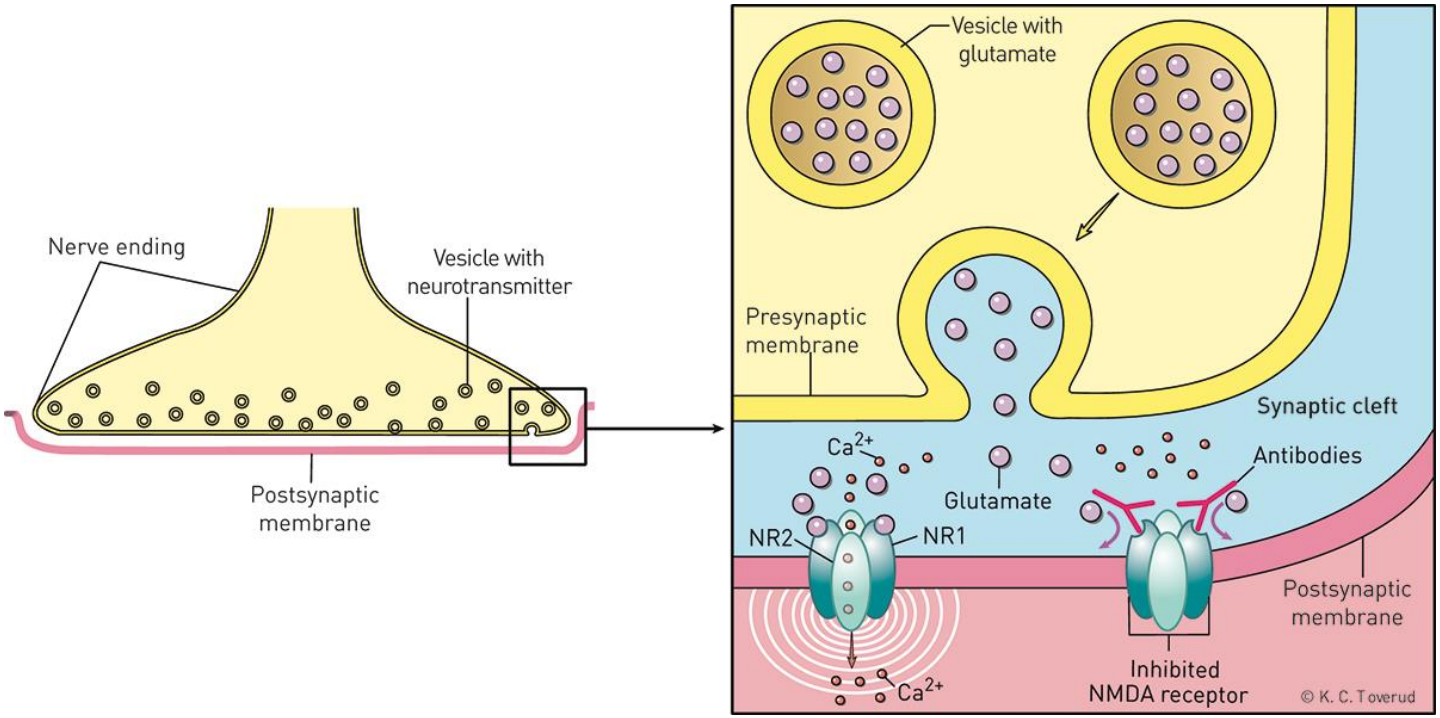


N-Methyl-D-Asparate (NMDA)



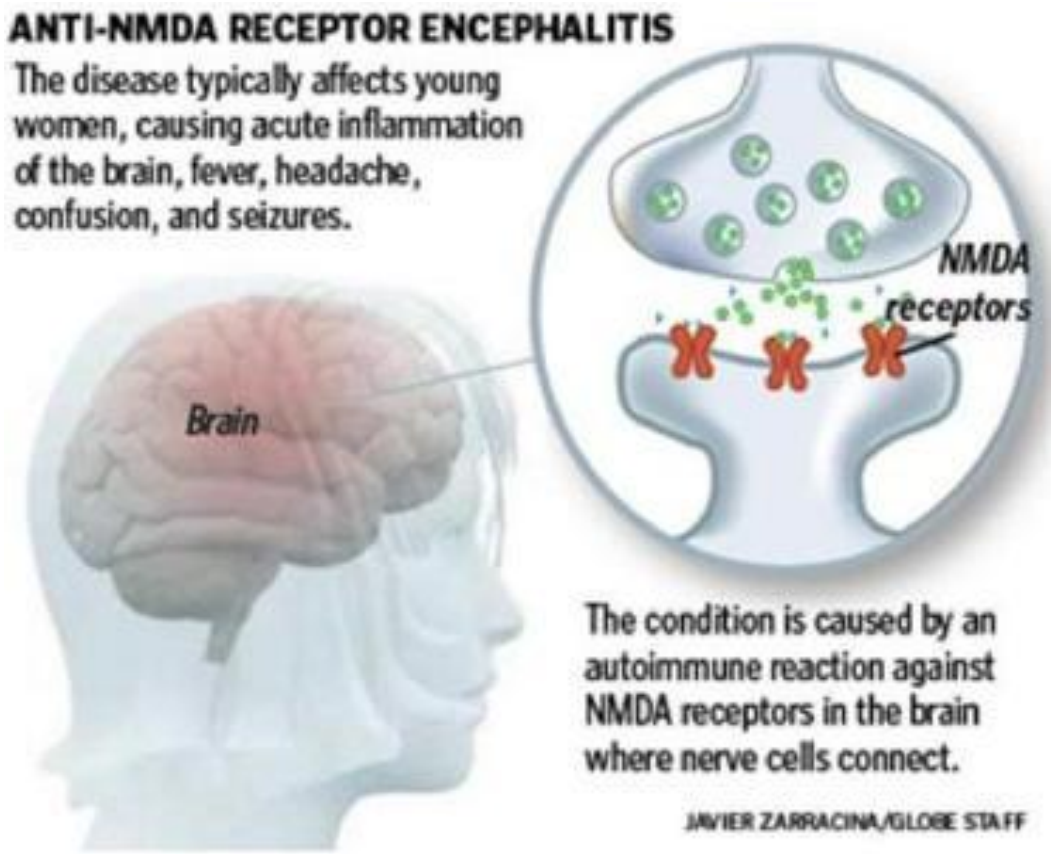


Mechanism and Pathophysiology



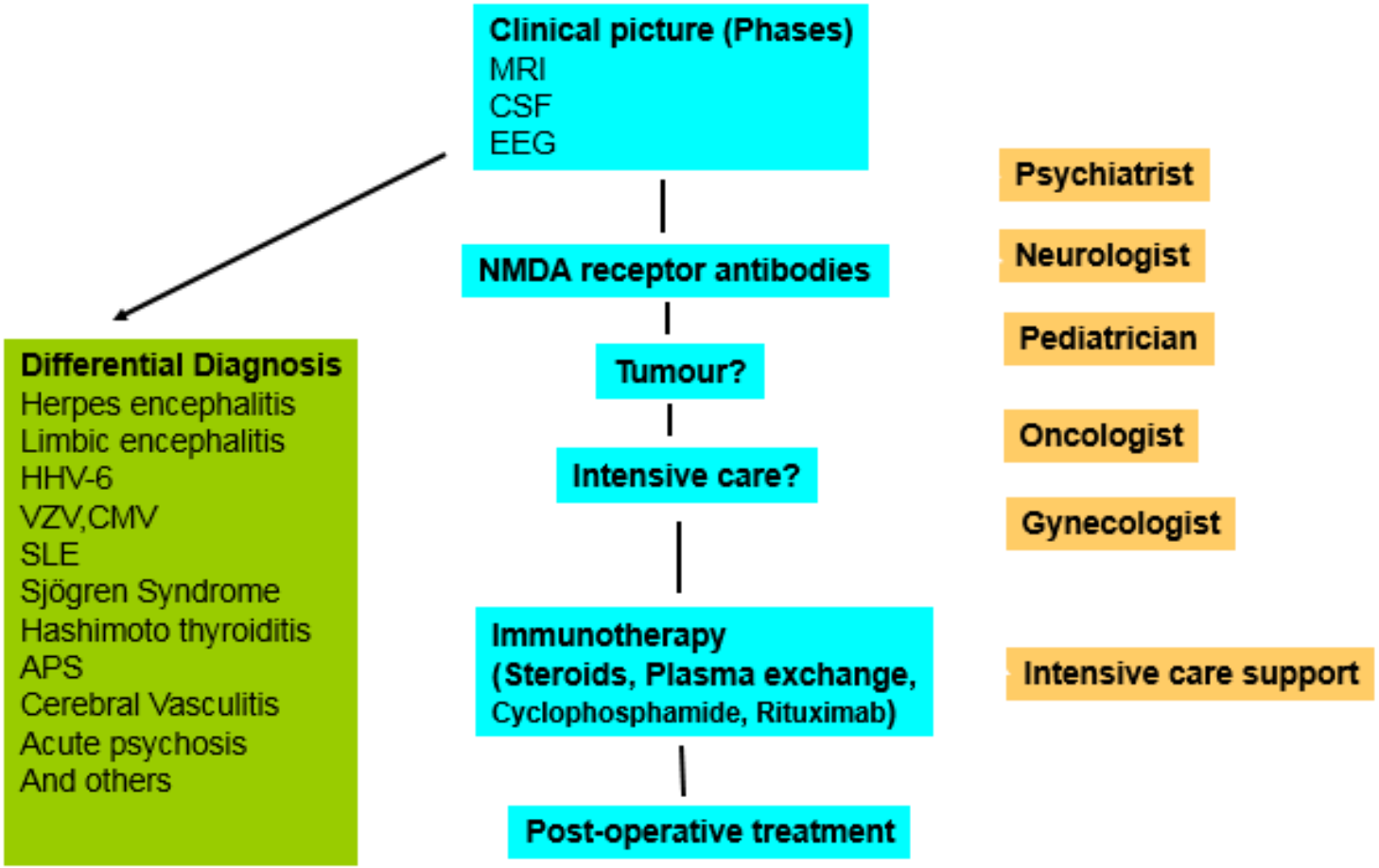
- 抗NMDA受體抗體 (抗谷氨酸受體抗體)
- 抗NMDA受體腦炎跟自體抗體攻擊NMDA受體的**NR1 subunit**有關

Mechanism and Pathophysiology



- 在抗NMDA受體腦炎中自體免疫能影響行為情緒記憶以及意識形態

診斷和治療



Source: H. Prüß, J. Dalmau, V. Arolt, KP. Wandinger , Nervenarzt 2010

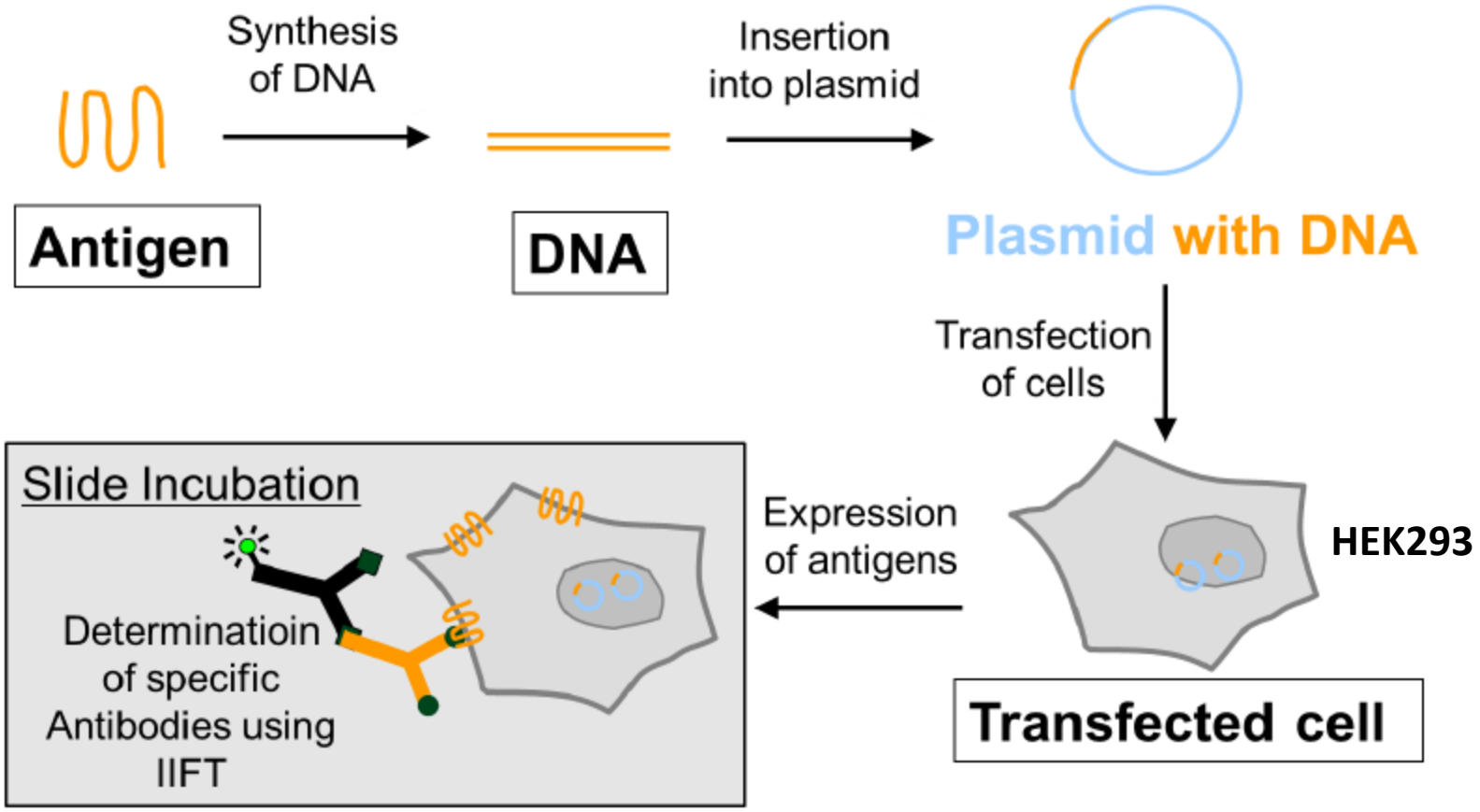
Laboratory Feature

- **The gold standard for neuronal surface Ab detection**
  - Method：Indirect Immunofluorescence Test (IIFT)
  - Specimen：Serum or CSF
  - Dilution factor：Serum at dilution of 10X or greater
    CSF at original titer to 10X
  - Incubated with recombinant HEK cells

Zuliani L et al . J Neurol Neurosurg Psychiatry (2012)

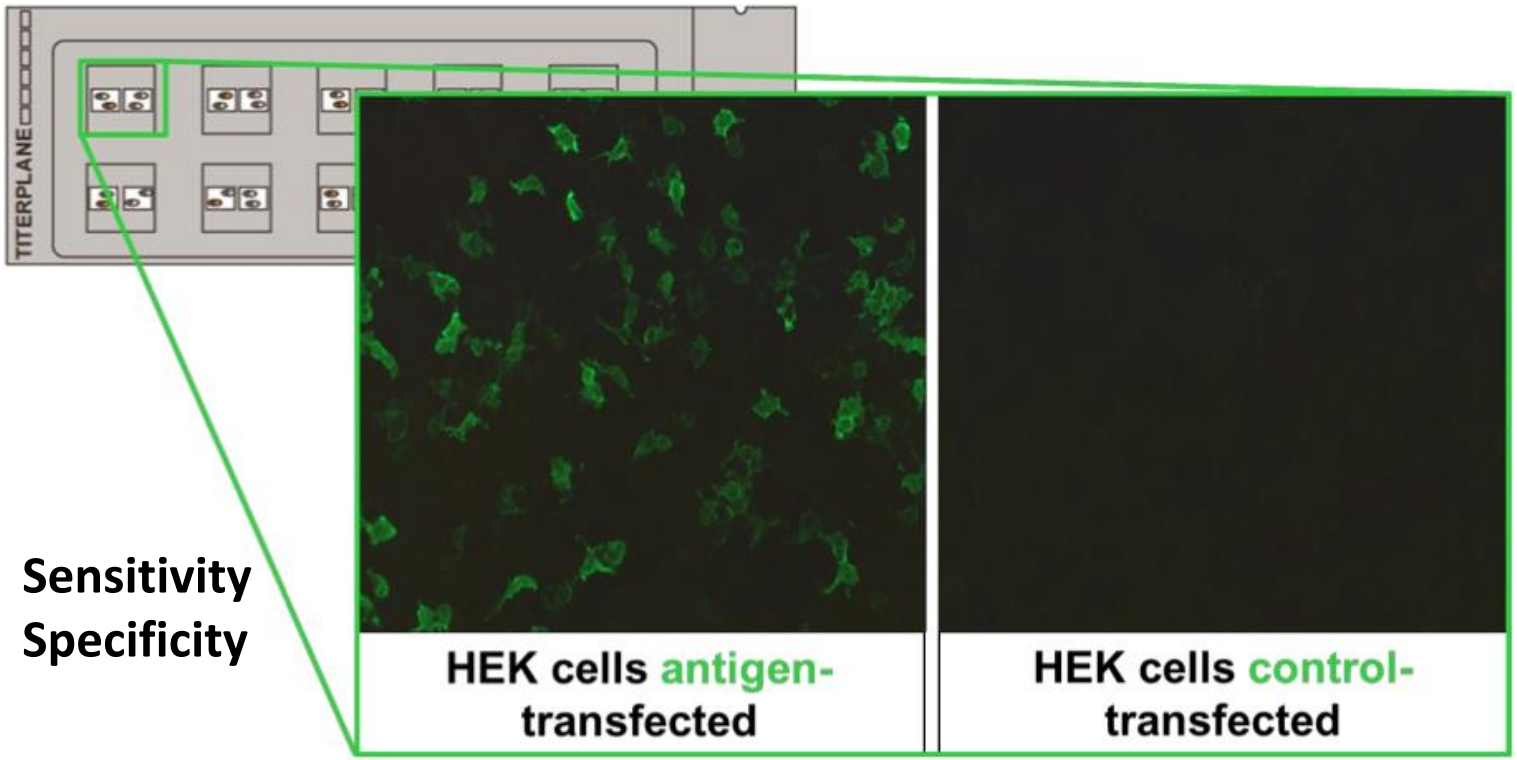
Laboratory Feature

- **The gold standard for neuronal surface Ab detection**



Laboratory Feature

- **Evaluation of recombinant cells (HEK)**

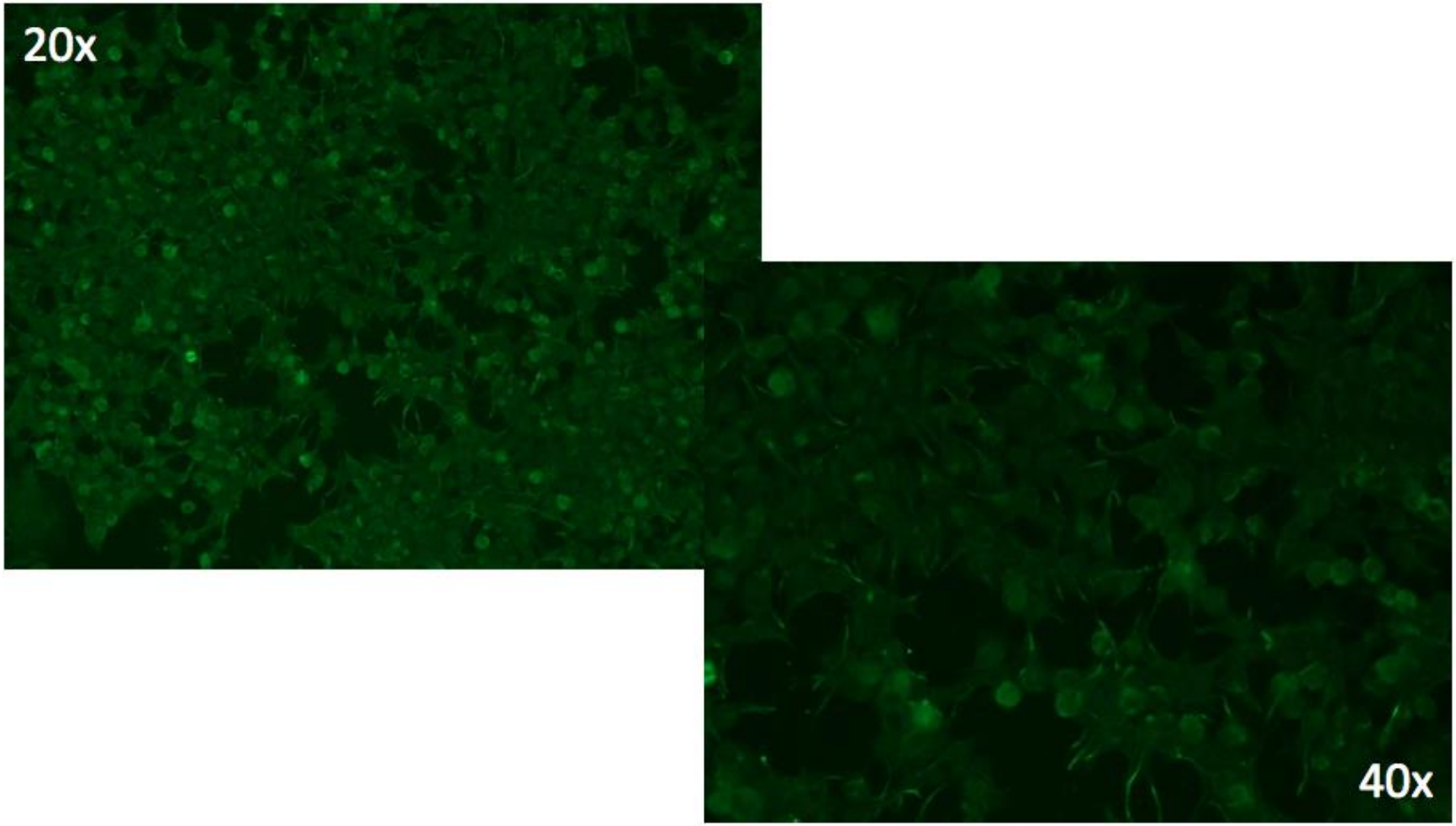


Sensitivity
Specificity

Recombinant Cell IFT (RC-IFT)

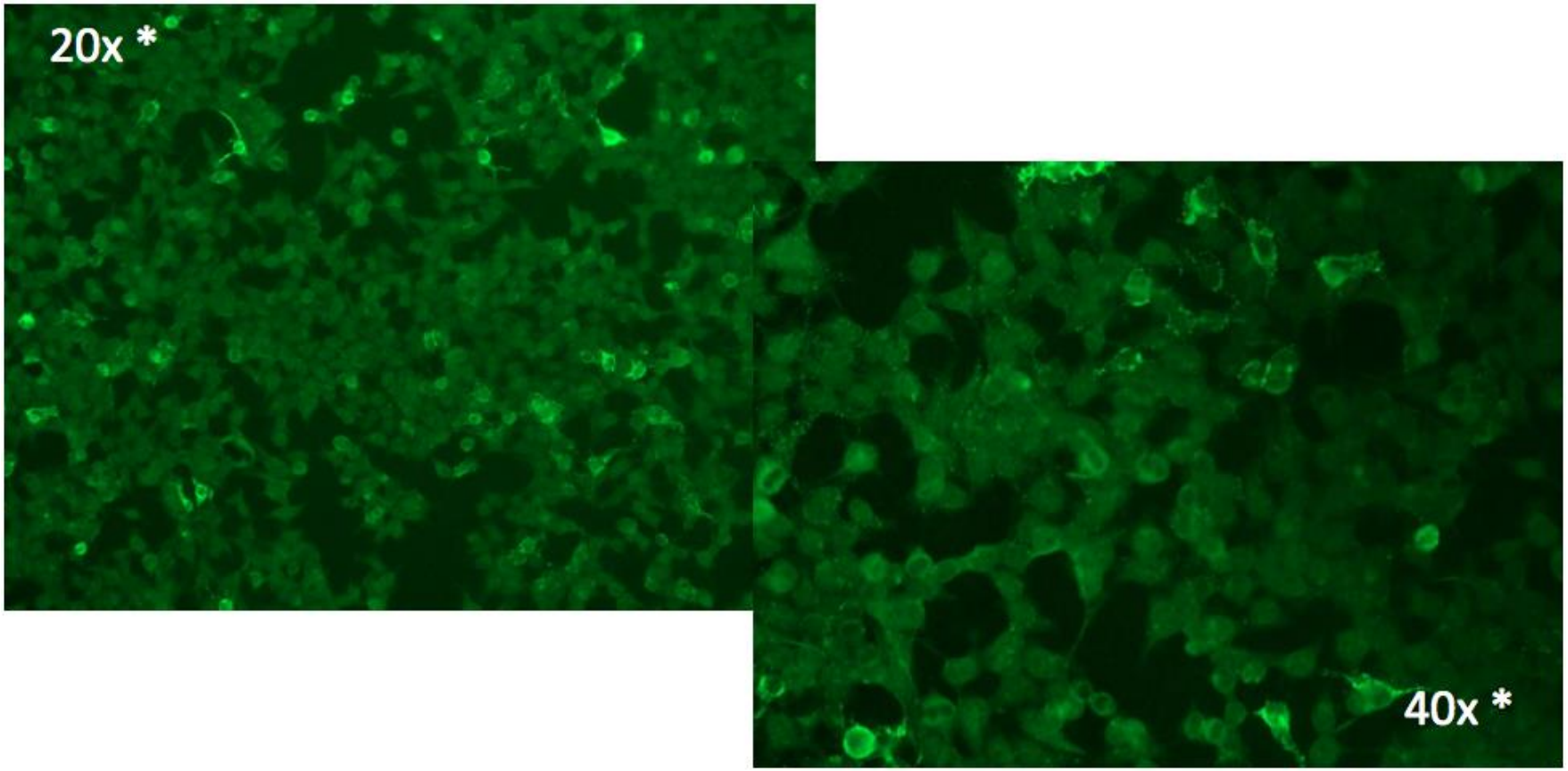
Laboratory Feature

Anti-NMDAR Negative pattern



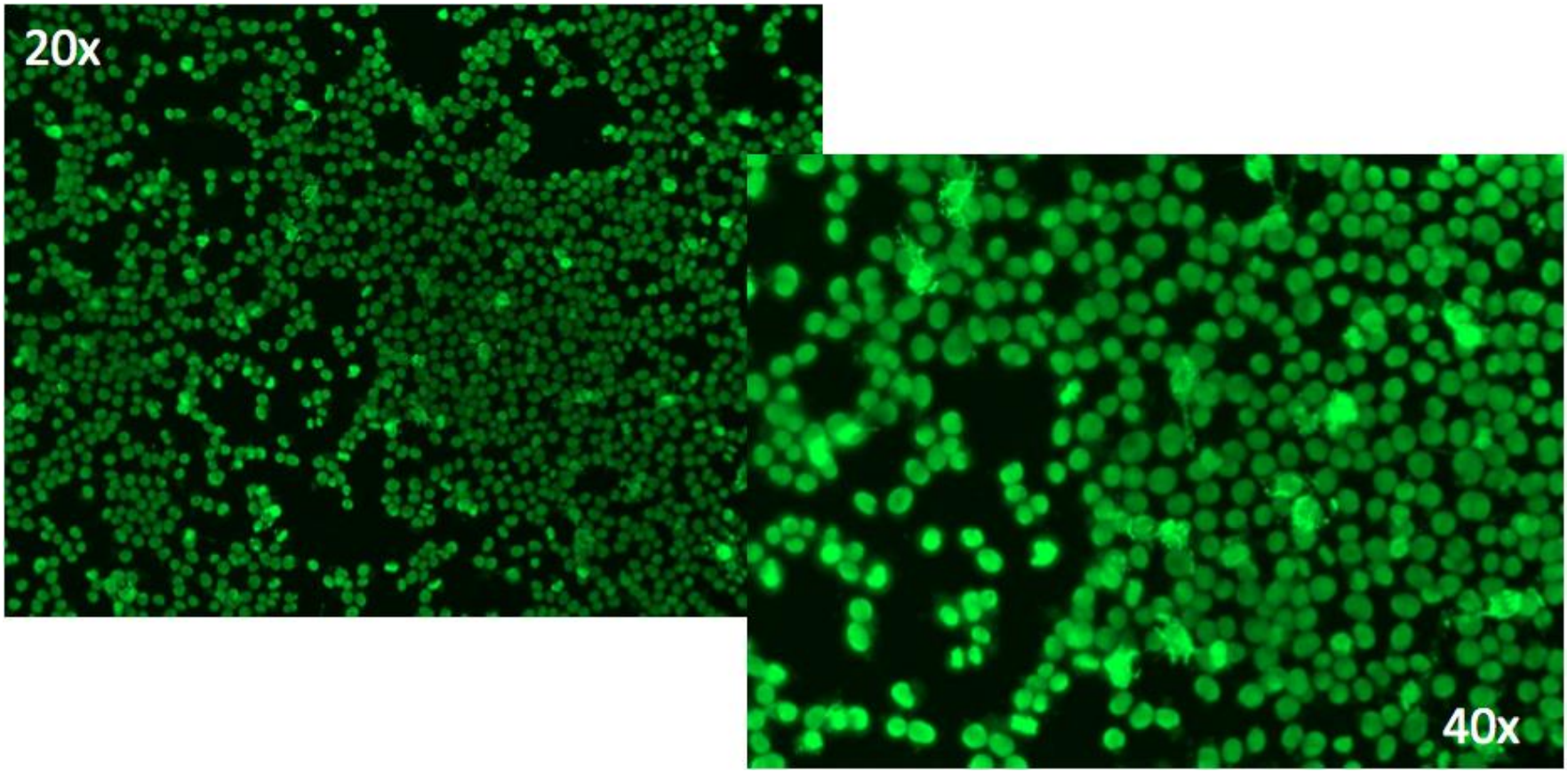
Laboratory Feature

Anti-NMDAR Positive pattern



Laboratory Feature

Anti-NMDAR and ANA positive



Treatment

- 治療抗NMDA受體腦炎的方法針對病患的病程有不同的醫療處置

- 第一線醫療處置：
  - 腫瘤的切除 (若其存在)
  - 類固醇藥物
  - 血漿置換 (血液透析)
  - 靜脈注射免疫球蛋白(IVIG)

- 第二線醫療處置：
  - 投以免疫抑制藥物
  - CellCept
  - Rituximab
  - Cytoxan

Recovery

- 復原之路緩慢且復原所需時間通常與疾病發展進程成反比
- 大多的病患能在疾病發生後開始治療的兩年內痊癒
- 病患要有好的預後通常包括幾個要素，疾病伴隨的腫瘤而發生，快速且確切的診斷以及積極的治療