

Sin
SOCIETÀ ITALIANA DI NEUROLOGIA

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**Rete Italiana della Tossina Botulinica
(RITB)**

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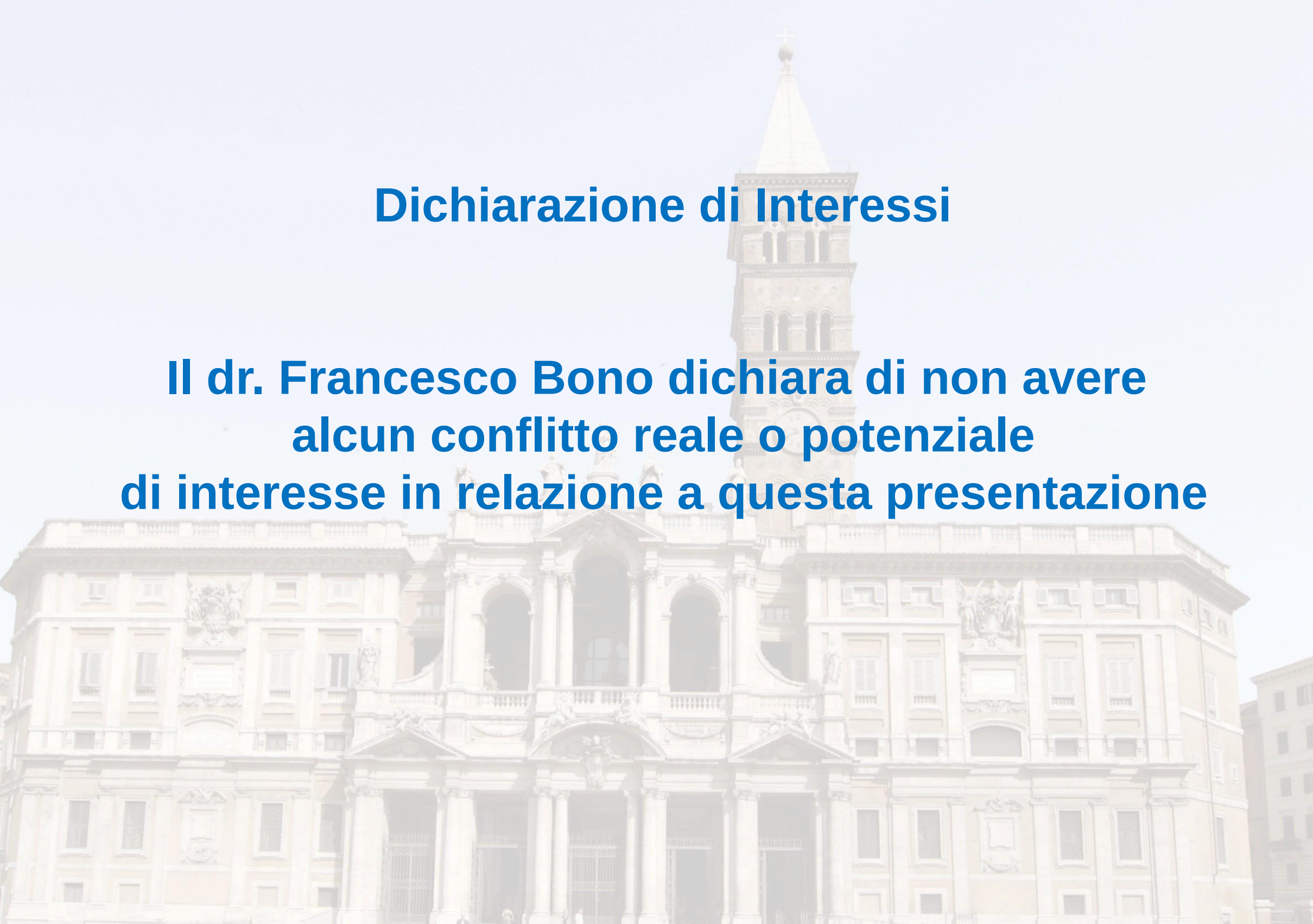
**LE DISTONIE CRANICHE
TRA ARTE E NEUROLOGIA**

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Dichiarazione di Interessi

**Il dr. Francesco Bono dichiara di non avere
alcun conflitto reale o potenziale
di interesse in relazione a questa presentazione**



La Distonia Cranica tra Arte e Neurologia: la Storia

XVI secolo (1558)

Peter Brueghel il vecchio dipinge “De Gaper”: la prima descrizione artistica



1910

Henri Meige descrive blefarospasmo/“spasmo facciale mediano”: la prima descrizione medica



1925

Jean A. Sicard usa il termine “para spasmo facciale bilaterale” per blefarospasmo/“spasmo facciale”



1972

George Paulson usa l’eponimo “Sindrome di Meige” per blefarospasmo e distonia oromandibolare



1976

Charles D Marsden descrive la “Sindrome di Brueghel”: blefarospasmo e distonia oromandibolare

Distonia cranica: la prima descrizione medica

Sindrome di Meige ?



Henri Meige. Le convulsions de la face: une forme clinique de convulsion faciale, bilaterale et mediane. Rev Neurol (Paris) 10 (1910) 437-443

Descrive 10 pazienti con involontaria chiusura delle palpebre

- 9 pazienti con isolato blefarospasmo
- 1 solo paziente con spasmo dei muscoli facciali inferiori
- “Spasmo facciale mediano”
- Lo considera un disturbo non organico
- Gli spasmi estesi alle regioni vicine possono essere causati da una lesione bulbo-pontina

Distonia cranica: la prima descrizione medica

Descrive il blefarospasmo ed i movimenti involontari dei muscoli facciali inferiori e masticatori



Horatio C. Wood, fr

“The contraction is tonic, causing a complete closure of the eye, and consequent blindness. This is accompanied by innumerable bizarre grimaces, due to the efforts of the antagonistic muscles to overcome the force which is closing the lids.”

Horatio C Wood. Nervous Diseases and their diagnosis: A treatise upon the phenomena produced by diseases of the nervous system. JB Lippincott, Philadelphia, 1887

La distonia cranica tra arte e neurologia la sindrome di Brueghel



“sindrome blefarospasmo-distonia oromandibolare”

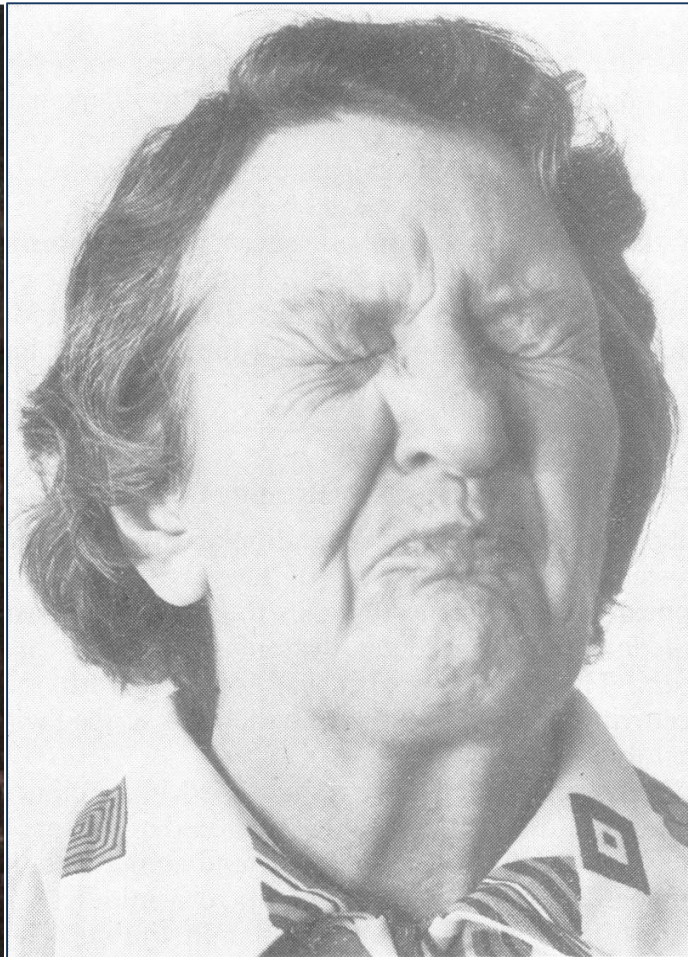
- Blefarospasmo 13 pazienti
- Distonia Oromandibolare 9 pazienti
- Blefarospasmo con Distonia Oromandibolare 17 pazienti
- “Adult-onset dystonia
due to an abnormality of extrapyramidal function”
- Usa il termine sindrome di Brueghel

•C. D. Marsden. Blepharospasm-ormandibular dystonia syndrome (Brueghel’s syndrome).
J Neurol Neurosurg Psychiatry 1976;39:1204–9.

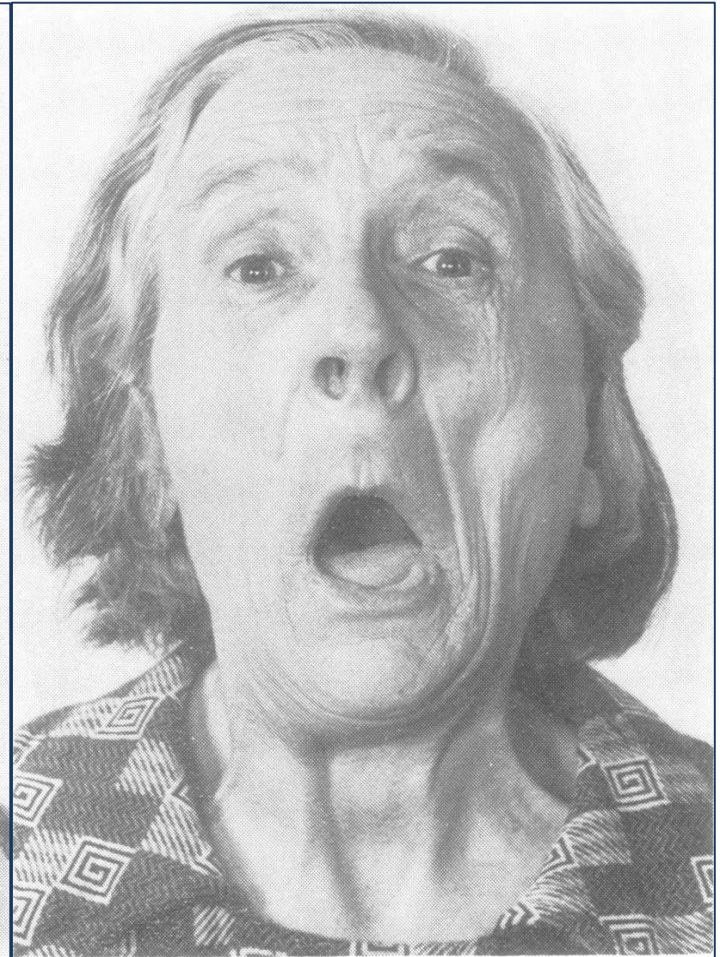
La distonia cranica: la prima descrizione artistica ?



"De Gaper"
Pieter Brueghel, The elder (c. 1558)
Musée Royaux des Beaux-Arts
Bruxelles.



C. D. Marsden, 1976:
" I am grateful to Dr R. E. Kelly
for pointing out that Pieter Brueghel,
the elder, recognised the syndrome"



Parkes et al. have suggested
that "it is simply the portrait
of a man jawning".



La Dystonia Cranica tra Arte e Neurologia

***A Man With Eyes Shut Tight*
Giovanni Agostino da Lodi
(c. 1506)**

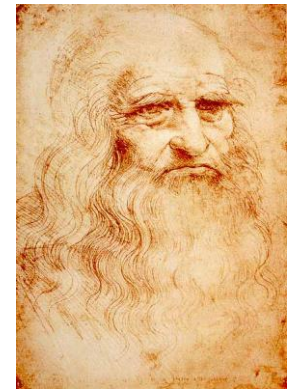
inv. 1895, 0915.481.

British Museum, Londra, UK

**Un pittore attivo dal 1495 al
1519 appartenente al gruppo
dei Leonardeschi.
“Pseudo Boccaccino”**



**Malaguzzi Valeri, 1912
Franco Moro, 1989
Giulio Bora, 1998**



**Leonardeschi:
Un gruppo di artisti allievi
di Leonardo da Vinci in
terra lombarda (diretti o
indiretti), che nei primi tre
decenni del cinquecento
avevano diffuso lo stile
innovativo di Leonardo in
aree estranee al suo
passaggio (Venezia,
Svizzera, Italia centro
meridionale).**

La Prima Descrizione Artistica della Distonia Cranica

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Facial dystonia as depicted in art in the time of Leonardo da Vinci

We would like to draw your attention to the red-chalk drawing *A man with eyes shut tight* by Giovanni Agostino da Lodi, an Italian renaissance painter and draughtsman of the Leonardeschi¹ (a group of artists who worked in the studio or under the tutorage of Leonardo da Vinci). This drawing (figure A) is one of a series of red-chalk studies of heads, often mistaken as works by Leonardo da Vinci, which da Lodi created after being inspired by similar drawings by da Vinci.² This physiognomic study of a male head is animated by the intensity of facial expression: the curly haired man has his eyes shut tight and is grimacing. Careful observation can recognise a middle-aged man with involuntary contractions of the orbicularis oculi muscles causing forceful closure of his eyes, while involuntary contractions of the zygomatic and masticatory muscles are causing clenching of his jaw and a grimace or forced smile. Additionally, an involuntary contraction of the left platysma muscle is evident from submandibular skin folds. The larger right sternocleidomastoid muscle, compared with the left, suggests involvement of the neck muscles, producing a slight left rotation of the head.

The same facial expression can be seen in a patient with involuntary eyelid closure associated with involuntary movements of the lower facial and masticatory muscles, causing clenching of the jaw and a grimace (figure B). A slight rotation of the head to the right side can also be seen. This combination of blepharospasm and involuntary movements of the lower facial or masticatory muscles with involvement of the neck muscles is characteristic of cranial-cervical dystonia.³

The American neurologist Horatio C Wood first described cranial

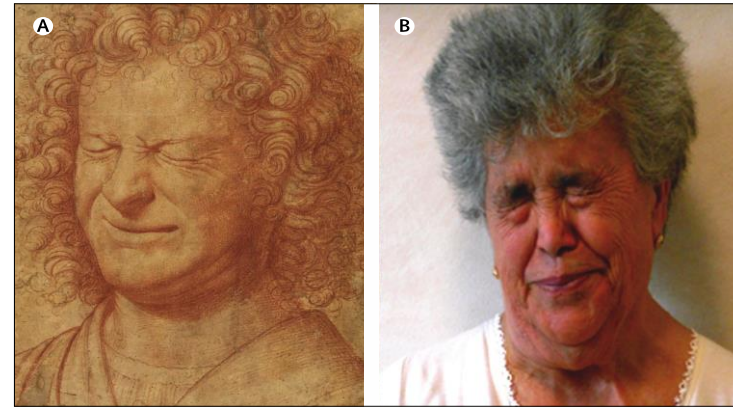


Figure: Facial dystonia in Renaissance art (A) and in 79-year-old patient (B)
Drawing (A) reproduced by permission of Trustees of the British Museum (London, UK).

dystonia in 1887, and noted that in facial and oromandibular dystonia "The contraction is tonic, causing a complete closure of the eye, and consequent blindness. This is accompanied by innumerable grimaces".⁴ In 1976, the English neurologist Charles David Marsden drew attention to the 16th century painting *De Gaper* by Pieter Brueghel the Elder, which depicts cranial dystonia.⁵ This painting shows the head of a man with his eyes partly closed and mouth wide open. Marsden⁵ suggested that the image depicted an individual with blepharospasm and oromandibular dystonia, and he used the eponym Brueghel's syndrome, but some authors⁶ have suggested that the portrait is simply of a man yawning.

Eponymic terms are now deemed inappropriate, and the term cranial-cervical dystonia should be used for the combination of blepharospasm and dystonia of other muscles of the head and neck.^{3,7}

On the basis of the clinical features of the individual in the drawing by Giovanni Agostino da Lodi, we think that this physiognomic study of a male head represents an artistic description of cranial-cervical dystonia. Importantly, this drawing is dated to the early 16th century (1500–06).^{1,2,8}

Thus, well before the painting by Pieter Brueghel the Elder (1558),⁵ a pupil of Leonardo da Vinci might have made the first artistic description of cranial-cervical dystonia.

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Francesco Bono

La Distonia Cranica tra Arte e Neurologia: una storia riscritta

XVI secolo (1500-06)

Giovanni Agostino da Lodi disegna *"A Man With Eyes Shut Tight"*
La prima descrizione artistica della distonia cranica



1887

Horatio C. Wood descrive il blefarospasmo ed i movimenti involontari dei muscoli facciali inferiori
La prima descrizione medica della distonia cranica



1910

Henri Meige descrive il blefarospasmo come "spasmo facciale mediano"



1972

G. Paulson riconosce che il blefarospasmo e la distonia oromandibolare hanno una patogenesi comune



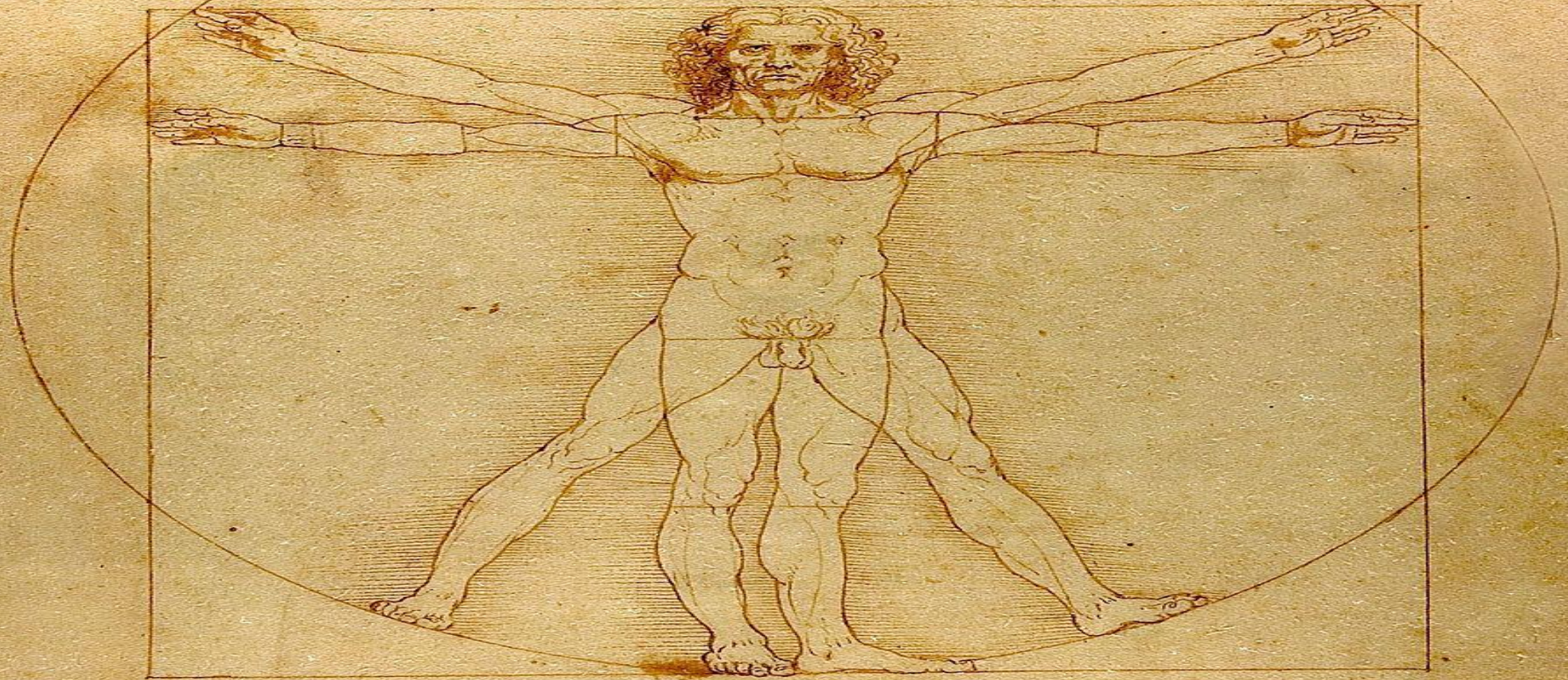
1976

C. D. Marsden descrive la sindrome blefarospasmo-distonia oromandibolare come distonia cranica

120

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**"LA MEDICINA
E' UNA COMBINAZIONE TRA ARTE E SCIENZA"**



Grazie per l'attenzione