Abuse of the synthetic amphetamine deriva-
tive, 'ecstasy' (3,4-methylenedioxyamphetamine, 
MDMA) causes convulsions and sudden cardiac death, hyperpyrexia, 
acute renal failure, psychosis, and cerebral haemorrhage. We report a previously 
unrecognised, serious neurological complica-
tion.

After ingestion of a single tablet of 
MDMA at a dance party, a 22 year old 
woman spent eight hours dancing without 
drinking any fluids. Twelve hours after tak-
ing the MDMA she developed throbbing 
headache, nausea and photophobia, 
followed by visual fortification spectra, expres-
sive dysphoria and right hemisensory loss. 
After 2 hours the sensory loss and dysphasia 
resolved, but she remained confused and 
afebrile with no history of cerebral 
venous sinus thrombosis. Three months later 
including lupus anticoagulant, and 
antiphospholipid antibodies, and antineu-
rophil cytoplasmic antibodies. 

To investigate the possibility that facial 
haemorrhage results from localised auto-
nomic dysfunction, sympathetic and sino-
atrial function were performed on the 
proband. Both sides of the face were tested, 
the clinically normal side acting as an inter-
natural. The accuracy of pupillary reactions was investigated using the 
following eye drops: 2-5% metha-
choline, 4% cocaine, and 1 in 1000 adrena-
line. All reactions were as expected for the 
normally innervated pupil with no evidence 
of parasympathetic or sympathetic denerva-
tion (central or peripheral). Lacration 
was investigated by the Schirmer tear test; 
this was found to be symmetrical and within 
normal limits. Solumedrol function was 
investigated by a standard sweat test with 
quinzinarin staining; no asymmetry in 
response was detected. No evidence of pos-
tural hypotension was found and there was 
no angulation in heart rate following oral 
carotid sinus massage or ocular pressure.

Hemifacial atrophy is a disorder of uncer-
tain aetiology. Because of the diversity of 
clinical features seen in various cases, the 
unifying entity of the syndrome has been ques-
tioned, prompting the suggestion of possi-
bile aetiological heterogeneity.

The occasional familial occurrence of 
HFA suggests the operation of a genetic 
factor in the causation of some cases. Reviewing such reports, McKusick3 came 
to the conclusion that HFA was inherited as an 
autosomal dominant condition with incomplete penetrance; a single case of  
scleroderma was reported and the importance of facial 
innervation was questioned.2 The 
author reasoned that the discovery of a 
patient with HFA allowed us to address 
some of these controversies.

A 23 year old left handed white male 
complained of progressive wasting of the 
right side of his face. He was the second 
born of identical twins, delivered per vaginam without instrumental assistance. 
His developmental milestones 
ocurred at appropriate times. He first 
noticed thinning of the face at age 17, 
particular around the chin and later around 
the malar prominence. This was con-
formed by contemporaneous photographs. 
In retrospect, he could recall being bitten 
on the chin during a fight at age 14 but this 
did not result in bleeding or bruising. There 
was no history of facial numbness, 
migraine, seizures, or symptoms of bulbar 
 palsies, and family history was negative 
for similar conditions.

Clinical examination revealed wasting of 
subcutaneous tissues on the right side of 
the face, including platysma. There was no neu-
rological deficit in the cranial nerves, in 
particular trigeminal nerve function was 
normal; the limbs were symmetrical and 
neurologically intact. Thorough inspection 
of the skin showed no stigmata of sclero-
derma. Eyebrows and moustache were 
symmetrical with no achromia or hyperchromia, and no appreciable difference in facial sur-
face temperature or skin color was detected. 
The patient's right handed brother, the first born twin, was also exam-
ined and had neither facial nor limb hemi-
trophy, neurological and dermatological examination was also normal.

DNA fingerprints were prepared from 
the blood of the proband and his twin 
(Celmark Diagnostics, Abingdon, UK) and 
identified identical bands, thus confirming 
monozygoty.

Serology proved negative for antinuclear 
antibodies, anti double-stranded DNA anti-
bodies, anti-centromere antibodies, anti-
ScC170, anti-Ro, anti-La, and anti-SS-A, 
extractable nuclear antigens, and anti-neu-
rophil cytoplasmic antibodies.

Some observations on the aetiology 
of progressive hemifacial atrophy 
("Parry-Romberg syndrome")

Although clinical reports of progressive 
facial atrophy (HFA) have appeared for 
over 150 years,1 the causes of the con-
dition remain unknown. Various theories, 
some underpinned with experimental evi-
dence, have been advanced, including a 
genetically determined disturbance of facial 
development, possibly inherited as an auto-
osomal dominant condition with incomplete 
penetrance; a localisation of scleroderma, or 
a disturbance of facial autonomic innervation, 
either central or peripheral. The discovery of a further 
patient with HFA allowed us to address 
some of these controversies.

A 23 year old left handed white male 
complained of progressive wasting of the 
right side of his face. He was the second 
born of identical twins, delivered per vaginam without instrumental assistance. 
His developmental milestones 
ocurred at appropriate times. He first 
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face temperature or skin color was detected. 
The patient's right handed brother, the first born twin, was also exam-
ined and had neither facial nor limb hemi-
trophy, neurological and dermatological examination was also normal.

In the control group, a similar frequency of abnormalities was found.7-8 The presence of these 
findings was associated with a history of 
MDMA use in the proband and her twin sister. 
There was no history of primary antiphospholipid syndrome, nor of the clinical features of 
neuropsychiatric MDNA associated with 
MDMA.8 However, the proband's maternal 
grandfather had a history of cardiomyopathy, 
and her maternal grandmother had died 
in cerebral haemorrhage. The proband's 
maternal grandmother had died in cerebral 
haemorrhage. The proband's maternal 
grandfather had a history of cardiomyopathy, 
and her maternal grandmother had died 
in cerebral haemorrhage. The proband's 
maternal grandmother had died in cerebral 
haemorrhage.
Cerebral venous sinus thrombosis induced by 'ecstasy'.

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