Clinical and Experimental Scleroderma 2014

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Systemic sclerosis (SSc) still presents
a major diagnostic and management
problem for patients and their physi-
cians, but despite this, the past twelve
months have witnessed some important
events in the field. In late 2013, the new
EULAR/ACR classification criteria
were published, an event which facili-
tates entry of patients into therapeutic
trials and observational cohorts at an
earlier stage of their disease. The Third
Systemic Sclerosis World Congress
gathered together physicians, health
professionals and patients in a compre-
hensive programme in Rome in Febru-
ary 2014 and the World Scleroderma
Foundation recently deepened its col-
laboration with EUSTAR to further SS-
related research and teaching globally.

An increasing interest in SSc has re-
sulted in a gratifyingly high number
of quality original publications in the
field, many of which are contained in
this edition of Clinical and Experimen-
tal Scleroderma. Basic research aspects
include the finding of antiphospholipid
antibodies in many of those with SSc,
and dysfunctional arteriovenous anas-
tomoses in the hands of many patients,
emphasising the importance of the vas-
cular component of SSc pathophysi-
ology. Also, the fibrotic component
continues to be seen as critical, as evi-
denced by the finding of overexpres-
sion of Toll-like receptors (including
TLR 5 and 10) in fibroblasts of SSc
patients.

For the clinician, useful data have been
reported regarding everyday manage-
ment of SSc patients, such as the im-
portance of Vit D and calcium supple-
ments for preventing fragility fractures,
the risk of Mg and Ca deficiency with
continuous high dose PPI use in some
cases, and practical algorithms for man-
aging the difficult GIT aspects of SSc.

In addition, recent data support the in-
creasing awareness that early interven-
tion is advisable, since much of the tis-

tue damage such as joint contractures
occurs in the first five years or so. Re-
lated to this is the early, simple, non-in-
vasive screening for potentially serious
complications such as pulmonary artery
hypertension, also covered by some pa-
pers here.

This theme of early detection and di-
agnosis and treatment of SSc has been
central to several public awareness pro-
grammes aimed at patients and primary
care physicians in close collaboration
with patient organisations.

The global trend of patient participation
in all aspects of clinical research is re-

flected also in this volume; refinements
in outcome measurements, such as
cross-cultural validation of established
instruments, and the measurement of
the impact of hand and face disability
on quality of life, all depend on close
patient co-operation in the form of pa-
tient reported outcomes (PRO).

Finally, on the treatment front, some en-
couraging but preliminary data are re-
ported regarding drugs such as endothe-
lin receptor blockers, tyrosine kinase
inhibitors and anti B cell monoclonal
antibodies, but clearly larger controlled
studies are required. The publication of
the ASTIS trial in JAMA this year has
given hope that a truly disease-modify-
ing strategy may be possible with SSc,
though currently this comes at the price
of significant toxicity. One of these new
developments, the emergence of new
autoimmunity in the form of ANCA
positive vasculitis, is reported here.

The reality-based hope is that through
the increased dissemination of data
and experience among colleagues in
academia, clinical practice and industry,
together with patients and regulators,
the future for patients and their families
with SSc will be brighter. This volume
represents such a sharing of practical
ideas on which to base this hope.

Competing interests: none declared.