AUDIBLE

HYPERACUSIS IN CHILDREN

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Introduction

Hyperacusis describes intolerance to certain (not necessarily loud) sounds by normally – hearing individuals.

The prevalence in the general adult population is not known but thought to be less common than in childhood. Research by Defeating deafness suggested that 6% of the children population may suffer over sensitivity to noise (2).

Hyperacusis can result from peripheral auditory pathologies for example; Bells palsy, Menieres disease, perilymph fistula and the recruitment associated with cochlear outer hair cells damage.

Central hyperacusis, however, is particularly prevalent (90%) in Williams Syndrome. Also, as many as 40% of children with communication disorders and autistic features are sensitive to noise.

Several other conditions have been reported to co-occur with central hyperacusis: migraine, depression, pyridoxine deficiency, benzodiazepine dependence, musicogenic epilepsy, Tay- Sach's disease, post-traumatic stress disorder, chronic fatigue syndrome and Lyme's encephalopathy.

Marriage and Barnes (5) postulated that 5-HT (Serotonin) dysfunction is a probable cause for the phenomenon of central hyperacusis. However, little research has been done in this area and therefore little is known about it.

In the last few years, it appeared that more children were being referred to Audiology services with hyperacusis. As a matter of curiosity and with the observed parallel increase in the number of children diagnosed with autism and ADHD, cases with hypersensitivity to sounds seen by the author in the last two years were collected and analysed.

Results

The total number of children with hyperacusis seen in the period Jan. 2000 to Nov. 2001 was 17 (M = 10, F = 7).

The majority of them (9) were referred because of concern about their hypersensitivity to noise. In four children, parents highlighted the abnormal reaction to noise during a follow-up review assessment of hearing. In the remaining 4, the main concern was regarding hearing and / or speech.

Referrals of children with hyperacusis were mainly by the family health visitors (**Table 1**). This is probably due to the fact that the majority (10) were pre-school age children (see **Figure**). The teenager (15 yr. old girl) suffered with photophobia as well as hyperacusis and diagnosed later to have post-viral chronic fatigue syndrome.

The majority (12) had had no significant hearing loss. One child (8 yr. old boy) was known to have unilateral SNHL following meningococcal meningitis at the age of 2 years. Four children had very mild conductive losses, in two, unilaterally.

An underlying pathology with a history of an insult to the brain was identifiable in all cases but 5. **Table 2** gives details of the individual cases.

Prematurity with consequent known disabilities i.e. cerebral palsy and / or hydrocephalus and / or epilepsy was present in 3 cases.

Meningococcal meningitis / septicaemia was present in another 3 cases.

The teenage girl developed chronic fatigue syndrome following mumps' meningitis.

In three cases, there was a history of maternal drug abuse during pregnancy. One of them who suffered neonatal withdrawal symptoms had had no other known pathology. In a second case with no other identifiable pathology, both parents abused Cannabis before and during pregnancy. The third case had had a history of meningitis at the age of 18 months in addition to the parental drug abuse.

The three children with communication disorders were on the mild end of the autistic spectrums. Two of them presented with hyperacusis before the diagnoses were made. The third case was a known Aspergers syndrome and her mother expressed concern about hypersensitivity to certain noises during a follow up assessment.

In six children there were associated behavioural problems and short concentration span. Interestingly, in all the six, there was an underlying pathology to the hyperacusis (**Table 3**).

Discussion

There is very little known about the prevalence, aetiology and management of hyperacusis in children.

The prevalence may be higher than perceived and on the increase. A specific enquiry about the presence of hyperacusis in the conditions known to be associated with this phenomenon might prove useful in elucidating the true prevalence.

This study identified three more conditions not recognised in the past to be associated with central hyperacusis.

The fact that three of the children had parents with positive **drug abuse** during pregnancy is very significant. The damage to the developing foetal brain that can be caused by gestational drug abuse is not well known. Central

hyperacusis in young pre-school children with no known pathology should raise suspicion of possible drug abuse during the early weeks of gestation, particularly with soft drugs like cannabis.

Monitoring of the incidence of central hyperacusis in young children and any observed increase in prevalence might reflect the high prevalence of drug abuse in the general population.

The possible link between central hyperacusis and early insult on foetal brain by cannabis or other drugs is a sensitive area to investigate. However, possible future re- classification of cannabis by the Home Secretary may prove very helpful!

The sequelae of **bacterial meningitis** including hearing loss is well known. However, hyperacusis was not mentioned among the many complications listed in a recent large study among 1584 five years old survivals in England and Wales (1).

The teenage girl with chronic fatigue syndrome (CFS), who had had both photophobia and hyperacusis suffered viral meningitis with positive identification of Mumps virus as the causative agent. It is well known that patients with CFS complain of hyperacusis. Whether the central hyperacusis in this patient was due to the post viral CFS or a direct consequence of the Mumps' meningitis is a theoretical question that would probably be difficult to resolve.

Similarly, **prematurity** with its many complications including hearing loss was not among the list of conditions associated with central hyperacusis (5).

It is important to emphasize that all cases of meningitis and prematurity included in this study (except one case with unilateral SNHL) had no significant hearing loss. Interestingly, the boy with unilateral SNHL following meningitis at the early age of 2 years started complaining of hypersensitivity to noise at the age of 8 years and it was bilateral. It coincided with the onset of behavioural problems. The unilateral SNHL was confirmed at an earlier age of 3.5 years.

The association between hyperacusis and behavioural problems / ADHD may have a common pathological background.

A decrease in forebrain 5-HT (Serotonin) activity was suggested as the most likely underlying pathology causing central hyperacusis (5).

Pyridoxine (Vitamin B6) has been used to treat hyperacusis in patients with communication disorders and autistic features (3). It is a co-factor in the synthesis of 5-HT and GABA.

Carbamazepine (which modulates GABA) has been used successfully in the treatment of hyperacusis in Lyme's disease (6). Lyme's encephalopathy is thought to preferentially involve the frontal cortical and sub-cortical areas including the limbic structure, with radiological evidence of an abnormal auditory-limbic link.

Imbalance in serotonin level in the brain was also thought to be the probable cause for hyperactivity symptoms in ADHD. The administration of a selective serotonin reuptake inhibitor (SSRI) in "ADHD" mice seemed to mimic the action of methylphenidate (Ritalin) (4).

Can Ritalin alleviate the symptoms of hyperacusis in children with ADHD? More interestingly, can it alleviate central hyperacusis associated with other conditions? I think it is worth a trial.

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Source of referral	No.
Family Health Visitor	8
Paediatricians	3
School Nurse	1
Mother	1
Review (mother)	4

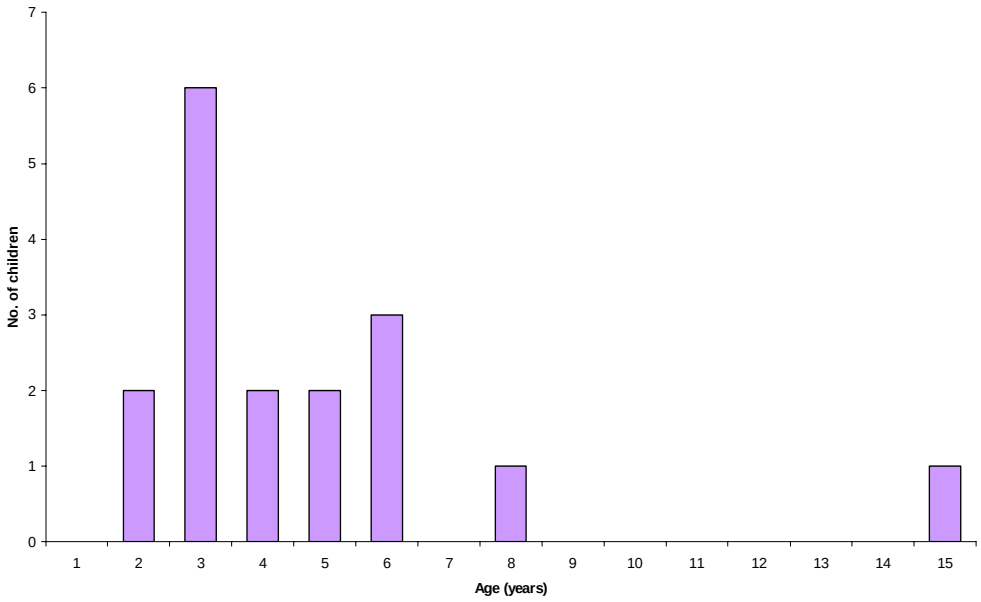
Table (1) shows that majority of children were referred by the family health visitor. In five children, the mother raised concern about her child's hypersensitivity to noise, 4 during a review assessment.

Conditions / Pathologies	No.
Prematurity & Cerebral palsy	1
Prematurity & Hydrocephalus	1
Prematurity, Cerebral palsy, Hydrocephalus & Epilepsy	1
Meningococcal meningitis	1
Meningococcal septicaemia	1
Meningococcal meningitis & Gestational drug abuse	1
Gestational drug abuse (withdrawal symptoms)	1
Gestational drug abuse	1
Chronic Fatigue Syndrome (Mumps meningitis)	1
Communication disorders / ASD	3
No known pathologies	5

Table (2) gives detail of the pathologies / conditions for all the 17 cases. Note that some cases have more than one underlying condition while in 5 cases there were no known pathology.

Pathology	No.
Meningitis	2
Prematurity & Hydrocephalus	1
Prematurity & Cerebral palsy	1
Communication disorder	2

Table (3) shows that in all the six children presented with hyperacusis and hyperactivity & / or short concentration span, there was an underlying pathology.



This figure shows the age distribution of the children

Guidelines for Medical Investigation of Bilateral Severe to Profound Permanent Deafness in Childhood

The British Association of Audiological Physicians (BAAP)
The British Association of Community Doctors in Audiology (BACDA)
Mac Ardle BM, Fonseca SJ.

Background

Advances in genetics, improvements in imaging techniques and changes in the epidemiology of childhood deafness have influenced how we investigate children with hearing impairment.

Clinical guidelines are an integral component of clinical governance (1) and practice may be improved by robust evaluation of guidelines (2). Guidelines are ' systematically developed statements to assist decisions about appropriate care for specific clinical circumstances' (3) based on systematic reviews of research literature. They are not intended to restrict clinical freedom, but practitioners are expected to use the recommendations as a basis for their practice. Local resources and the circumstances and preferences of individual patients also need to be taken into account.

The NDCS states that "Families must be offered the opportunity for their deaf child to have aetiological investigations. These investigations must be carried out in accordance with local protocols based on nationally agreed standards" (4).

A BAAP working group on evidence-based recommendations for the investigation of newly diagnosed children with bilateral severe and profound permanent hearing impairment developed a set of guidelines. These were agreed by BACDA in 2002 and can be viewed on the BAAP website www.baap.org.uk. These recommendations are relevant to all professionals in Audiology, deaf children and their families. Where possible recommendations are based on and linked to the evidence that supports them.

These guidelines will be reviewed in September 2002.

There are several reasons why it is important to investigate deafness:

1. To try and answer parents who ask, "Why is my child deaf?"
2. To identify and manage co-exisiting medical conditions such as Jervell and Lange-Nielsen syndrome, Alport's syndrome, Neurofibromatosis type 2, Ushers Syndrome and vestibular hypofunction.
3. To assist the family when making decisions about the most appropriate managenent strategy for their child as regards communication mode, educational placement and cochlear implantation.
4. To inform genetic counseling.
5. To inform epidemiological data.

Aims

The aim of these guidelines is to propose a rational approach to the medical investigation of bilateral severe to profound permanent deafness in children.

Subjects

All children with bilateral permanent hearing loss and thresholds over 70 dB in the better ear averaged across 500, 1000, 2000 and 4000Hz.

Guidelines for Good Practice

Level 1 investigations

Level 1 investigations should be considered for every child. Timing will depend on several factors, including the family's readiness to proceed with tests, availability of local test facilities and the child's ability to cooperate with tests.

- Paediatric history:
  - detailed history of pregnancy, delivery and postnatal period
  - developmental milestones including speech, language and motor milestones
  - pre and post natal exposure to noise
  - ototoxic medications
  - severe head injury
  - ear disease
  - meningitis
  - viral illness
  - immunisation status
- Family history of deafness or risk factors associated with hearing loss in first and second degree relatives
- Clinical Examination:
  - inspection and physical measurement of craniofacial region
  - examination of the neck, skin and nails, limbs, chest and abdomen
- Developmental assessment
- Family audiograms: on 1st degree relatives (5)
- Electrocardiography (ECG): for prolongation of the (corrected) QT interval (6)
- Ophthalmological assessment:
  - assessment of visual acuity and fundoscopy
  - discussion of electro-retinography with ophthalmologist if motor milestones are delayed (7, 8, 9, 10)

- Urine examination (labstix) for microscopic haematuria (11, 12, 13)
- Blood test for Connexin 26 mutation (14, 15, 16, 17)
- MRI of Internal Auditory Meati or CT Scan of Petrous Temporal Bone (18, 19, 20)

Level 2 investigations

Level 2 investigations will be indicated from history and clinical findings. As with level 1 investigations, timing will depend on the family's readiness to proceed with tests, availability of local test facilities and how well the child can cooperate with tests.

- Serology:
  - to exclude congenital infection
  - to include maternal stored (booking) serum
- Haematology and Biochemistry where clinically indicated
- Thyroid Tests:
  - family history of thyroid disease
  - goitre present
- Immunology Tests where clinically indicated
- Metabolic Screen on blood and urine: where clinically indicated
- Renal ultrasound:
  - if child has preauricular pits or sinuses, branchial cleft or cysts
  - Mondini defect on imaging
- Clinical photography
- Chromosomal studies:
  - history of developmental delay
  - dysmorphic features
- Referral to Clinical Geneticist especially if the parents are consanguineous
- Vestibular investigations

What next?

These guidelines will be reviewed by the working group* in September 2002. The revised version will be printed and distributed to clinicians. It is our intention to conduct a fully funded national audit of practice by BAAP and BACDA members in 2003-2004. Please send your comments to Dr B MacArdle or Dr S Fonseca.

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AUDITORY

British Society of Audiology

Report of Review Steering Group held on 13/11/02

Elaine English, John Irwin, Johanna Beyts, Ted Evans, Linda Luxton observing, with email contributions from David Baguley, Pauline Smith, Clive Sparkes

1. "Umbrella organization" or "Academy of audiology"?

A minority of e-mails urged us to "scrap everything - BSA and all the audiological societies - and start again" using the American Academy of Audiology as a potential model. The majority of email input and all of the steering group were not however convinced that this should be our immediate goal. The American Academy had made the mistake of being too exclusive and in the opinion of several, self-congratulatory. One of the strengths of the BSA was that it included members from ***all*** 10 Groups (BAAP, BAAS, BAAT, BACDA, BAEA, BATOD, BSHAA, BAO-HNS, BSHT, RCSLT). At least in the foreseeable future there will continue to be the need for specialist Groupings whether they are entirely separate as at present, or formally come under the umbrella of the BSA, or become amalgamated.

2. Executive officer

This appointment is seen to be crucial to the whole enterprise. An audiological professional was preferred to the employment of a professional administrator. Secondment was the preferred model. To begin with, secondment for say two days a week as part of an individual's personal development plan might be attainable.

3. Representation and committees

The principle of equal representation established at the Novartis meeting is recommended.

We propose a largish Council, acting as at present but providing policy guidelines for the executive officer.

Composition:

- One representative from each Group (10)
- 3 "National Representatives": i.e. from Scotland, Wales and Northern Ireland to ensure coverage of issues arising from devolution.
- Chairman, secretary, treasurer, vice-chairman
- Chairs of Education, Programmes, Standards and Professional Affairs committees.
- Executive ex-officio.
- 22 in all.
- All elected.

Each of the Education, Programmes, Standards and Professional Affairs Committee's should have 10 representatives from each of the constituent Groups (although they may need to coopt individuals to get the work done!)

In addition, Special Interest Groups would continue to be required as at present. Their Chairs would be at liberty to attend Council for specific items concerning them and annual reports would need to be submitted.

4. Unifying issues within the remit of the new Society

- Setting of standards in audiology
- Lobbying government. The BSA rather than the RNID should be the chief line to government!
- Professional Affairs: Construction of ideal skill mix models in audiology

Instead of a prescriptive clinical model which could not be expected to apply in all circumstances and locations, we preferred an ideal skill mix model(s) for the optimal quality of patient care and development of research and training on the lines of those proposed for MHAS, Screening and Balance initiatives and that currently being produced in Scotland (see www.phis.org.uk: Review of Audiological Needs). As part of this exercise the different professional Groups could be circulated with a questionnaire designed to elicit the perceived professional competences and limitations of each Group and therefore how patient needs can best be met. A Professional Affairs committee is proposed to take this on board.

- Training and Communication: including Annual Convention - with parallel sessions for the different Groups together with plenary sessions: introductory and high-level
- Accreditation
- Pay and conditions issues: it would be useful to have for example an annual meeting of representatives and unions involved in audiology so that the latter could be brought up to date on audiological issues and could inform audiologists what is being done on their behalf
- Collection and coordination of audiological data

5. Exclusions from the remit:

- Registration – must be by independent bodies eg GMC for the medics
- The few aspects of training that are carried out e.g. by the Royal Colleges
- Detailed pay and conditions

6. Management of differences of opinion

The Executive would need a "hotline" with key individuals in the constituent Groups on the style of the Confederation of Health Trust Executives

Much greater communication between Groups needs to be addressed. This could be greatly assisted by the Professional Affairs committee; by an Annual Convention bringing together all of the Groups; by a beefed-up *News* giving space or even issues for specialist interests together with a biennial report on professional issues from each Group

7. New name?

The email suggestion - *British Societies of Audiology* - has merit!

8. Next step

Proposals emerging from the Nottingham meeting should be rapidly disseminated to the *whole membership* of BSA and to the Groups.

Opinion could be tested by questionnaire or referendum.

AUDIOLOGY

British Society of Audiology

Report of Societies' Representatives Meeting, Nottingham 22/11/02

Feluki Ajayi (BAAS), Sue Archbold/David Hartley (BATOD), Brian Barney (BAEA) , Caroline Fraser (RCSLT), Roger Lewin (BSHAA), Eva Raglan (BAAP), Paul White (BAAT); Peggy Chalmers, Ted Evans, Linda Luxon observing, John Wilderspin chairing.

The meeting unanimously endorsed the following proposals on the grounds that they potentially could confer significant advantages to audiology and to the individual constituent Groups. In particular, we see enormous value in having an effective organization able to formulate policy and best practice in audiology, to lobby effectively, to link the disparate Groups more efficiently, and to encourage multidisciplinary training and professional development.

1. "Umbrella organization" or "Academy of audiology"?

A minority of e-mail input urged us to "scrap everything - BSA and all the audiological societies - and start again" using the American Academy of Audiology as a potential model. The majority of opinion received, and unanimously that of the Meeting, was not however convinced that this should be our immediate goal. The American Academy had made the mistake of being too exclusive and in the opinion of some, self-congratulatory. One of the strengths of the BSA was that it included members from ***all*** 10 Groups (BAAP, BAAS, BAAT, BACDA, BAEA, BATOD, BSHAA, BAO-HNS, BSHT, RCSLT). At least in the foreseeable future there will continue to be the need for specialist Groupings whether they are entirely separate as at present, or formally come under the umbrella of the new organization, or become amalgamated.

The new umbrella would exist in parallel to newly created organizations such as the Federation of Health Care Scientists, and be complementary (specific to audiology), rather than in competition.

2. Executive officer

This appointment is seen to be crucial to the whole enterprise.

Key issues to be settled would be job description; affordability; appointment process; relationship between Executive, Chair and Council (a possible model could be the relationship between a school head and the school Governing body).

We were divided on the two possibilities: an audiological professional or a professional administrator.

An audiological professional had the advantage of professional audiological prestige and knowledge, and economy (if secondment was possible even on a day or two a week basis as part of an individual's personal development plan).

A professional administrator should have the advantage of lobbying skills, but the cost full-time would be at least £50k pa.

3. Membership

Two categories are proposed, but these would need to be informed by the proposed Treasurers' Meeting (see below):-

- Affilated members: members of Groups affiliated to the new organization; voting rights through the Group.
- Full individual membership: full voting rights and receiving the Journal, for example.

4. Representation and committees

The principle of equal representation established at the Novartis meeting is upheld.

We propose a largish Council, acting as at present but providing policy guidelines for the Executive Officer.

Composition:

- One representative from each Group (10), each elected by their Group and thereby becoming full members. The tenure should be two years.
- 3 "National Representatives": ie from Scotland, Wales and Northern Ireland to ensure coverage of issues arising from devolution, each elected by full members in each Region. The tenure should be two years.
- Chairman and Vice-chairman (future Chair) need to be drawn from different Groups from nominations by Council (to encourage balance over time) and elected by the full members. A one year tenure is suggested, including Past Chair.
- Treasurer: elected by Council.
- Chairs of Education, Programmes, Standards and Professional Affairs committees, elected or appointed by Council to ensure broad representation. (Could some of these be by secondment on a one or two day a week basis?). Tenure: two years.
- Executive Officer ex-officio.
- 22 in all.

In addition, Special Interest Groups would continue to be required as at present. Their Chairs would be at liberty to attend Council for specific items concerning them and annual reports would need to be submitted.

There should be a quorum.

At the end of tenure of membership of Council, re-election would not be permitted within two years.

It was felt that an Honorary Secretary would not be necessary with the new administrative structure.

The Chairman, Vice-Chair, Treasurer and Past Chair would be the Trustees, and form an Executive/Steering Group

together with the Executive Officer.

Named alternates would be essential.

UKCOD, representing RNID, NDCS etc would be circulated with agendas and invited to participate as observers when necessary to ensure lay/user input.

Each of the Education, Programmes, Standards and Professional Affairs Committees should invite representatives from each of the constituent Groups (although they may need to co-opt specific individuals to get the work done!) Alternates would be essential. The remits of these Committees would need to be devised.

5. Unifying issues within the remit of the new Society

- Learned Society.
- Setting of standards in audiology.
- Lobbying government.
- Training and Communication: including Annual Convention - with parallel scientific sessions for the different Groups together with plenary sessions: introductory and high-level. This would subsume the educational needs of the constituent Groups without precluding Groups from convening their own meetings to discuss eg professional-specific affairs.
- Encouragement and coordination, by consultation, of multi-professional training, eg of degree courses, CPD etc.
- Collection and coordination of audiological service data.
- Professional Affairs: eg construction of ideal skill mix models in audiology; workforce issues.

(It is proposed that instead of a prescriptive clinical model which could not be expected to apply in all circumstances and locations, the new organization consider as a key issue an ideal skill mix model(s) for the optimal quality of patient care and development of research and training on the lines of those proposed for MHAS, Screening and Balance initiatives and that currently being produced in Scotland (see www.phis.org.uk: *Needs Assessment Report on NHS Audiological Services in Scotland*). As part of this exercise the different professional Groups could be circulated with a questionnaire designed to elicit the perceived professional competences and limitations of each Group and therefore how patient needs can best be met. A Professional Affairs committee is proposed to take this on board.)

6. Exclusions from the remit:

- Registration – must be by independent bodies eg GMC for the medics.
- The few aspects of training that are carried out eg by the Royal Colleges.
- Pay and conditions.

7. Finance

A meeting of Group Treasurers should be convened as soon as possible. For some Groups, a substantial subscription to the new central body would be out of the question, unless they were perceived to be receiving substantial value in return. A possible way forward might be affiliate and individual subscriptions to the central organization, the latter including the Journal, for example. A successful central Convention, economies of scale in administration and effective lobbying all should contribute to perceived value.

The expenses of the 10 Group representatives on Council might be funded by the Groups; Regional representatives and Officers, by the new organization.

The location of Office accommodation needs to be considered carefully.

The new organization should remain a Charity.

8. Communication

The Executive would need a "hotline" with key individuals in the constituent Groups with identified areas of expertise. The Executive Officer would circulate position papers and elicit or prepare draft proposals/responses. The onus would be on the Groups to respond to requests for advice/decisions - silence signifying assent.

Much greater communication between Groups needs to be addressed. This could be greatly assisted by the Professional Affairs committee; by an Annual Convention bringing together all of the Groups; by a beefed-up *News* giving space or even issues for specialist interests together with a biennial report on professional issues from each Group.

Responsibility for monitoring and maintenance of good communication could be the responsibility of the Vice-Chair.

9. New name

Confederation of British Audiology. Alternatives: *British Societies of Audiology; British Audiological Society*.

10. Next steps

- A crucial question is that of finance (see 7 above). The outcome of the Treasurers Meeting should be circulated immediately.
- The current proposals should be rapidly disseminated to the *whole membership* of BSA and to the Groups.
- The present group should meet on 14th February at 10am in Birmingham to consider the proposals in the light of the financial implications.
- BSA would need to hold a consultation meeting in 6th March at 11 am in London and an EGM say in April 2003.