

**STICKLER SYNDROME
SUPPORT GROUP
(SSSG)
Registered Charity: 1060421**

**STICKLER SYNDROME:
A CHILD IN YOUR CARE?
WHAT TEACHERS AND
YOUTH LEADERS
NEED TO KNOW**

**BY
WENDY HUGHES**

INFO 11 11/2004

TABLE OF CONTENTS

1.	INTRODUCTION	1
2.	WHAT IS STICKLER SYNDROME?	1
2.1.	EYES	1
2.2.	ORO-FACIAL	1
2.3.	HEARING.....	2
2.4.	JOINTS	2
3.	CARE OF A CHILD WITH STICKLER SYNDROME.....	2
3.1.	GENERAL.....	2
3.2.	CLASSROOM MANAGEMENT	2
3.3.	ABSENCES FROM SCHOOL.....	3
3.4.	FATIGUE	3
3.5.	VISION	3
3.6.	HEARING.....	4
3.7.	JOINTS	4
3.8.	PSYCHOLOGICAL	5
3.9.	PHYSICAL EDUCATION AND SWIMMING.....	5
3.10.	MEDICATION.....	5
3.11.	ESTABLISHING A HOME-SCHOOL PARTNERSHIP.....	6
3.12.	FURTHER EDUCATION	6
4.	ABOUT THE SSSG	6
5.	HOW YOU CAN REACH US	7

1. INTRODUCTION

The Stickler Syndrome Support Group is frequently contacted by teachers/youth workers asking about any special considerations for a 'Stickler' child within their care. This booklet has been prepared to give those working with children in the 4-16 age range a better understanding of the condition and some of the more common problems that have been experienced. It is hoped that this booklet will provide a broad base for information to enable a Stickler child to have a full and rewarding education and to help them to reach their full potential.

2. WHAT IS STICKLER SYNDROME?

Stickler syndrome is a progressive genetic connective tissue disorder. It can affect both sexes and all ethnic communities and is usually passed on from parent to child. However about 10% of cases can occur for the first time in a child, but this means that the child concerned then also has a 50% chance (same as any parent) of passing on the condition to off-spring.

It was first defined, in 1965, by Dr Gunnar B Stickler, at the Mayo Clinic in the USA and named Hereditary Progressive Arthro-Ophthalmopathy, known world-wide as Stickler syndrome.

The disorder is associated with problems relating to vision, oro-facial features, hearing, and problems with bones and joints.

2.1. EYES

- High myopia (short-sightedness). This is usually severe -8 dioptries or more, although some Stickler syndrome patients are not myopic.
- High risk of retinal detachments, which may affect both eyes. A detachment occurs when the retina and the inner wall of the eye separate from each other - just like wallpaper peeling from a wall. When this happens the picture that is sent to the brain will become patchy, or in some cases completely lost. Early diagnosis is **vital** if blindness is to be prevented.
- Stickler syndrome is the most common inherited cause of rhegmatogenous (tears and holes) retinal detachments in children, therefore regular check-ups with an ophthalmic surgeon is essential.
- Pre-senile cataracts.
- Glaucoma.

2.2. ORO-FACIAL

- Children with Stickler syndrome can have distinct features as babies - flat face with a small button nose and little or no nasal bridge, pronounced upper lip groove, and a small lower jaw. Appearance tends to improve with age.
- Cleft palate, submucous or high arched palate and/ or bifid uvula.
- Micrognathia - where the lower jaw is shorter than the other resulting in poor contact between the chewing surfaces of the upper and lower teeth. These symptoms are similar to those found in Pierre Robin sequence. About 30% of children diagnosed as having Pierre Robin sequence at birth, subsequently have the diagnosis changed to Stickler syndrome about the age of 6-7 years when other problems become more obvious.

2.3. HEARING

- Hearing loss may be sensorineural (caused by nerve damage and results in a decrease in the ability of the nerve endings in the inner ear to transmit signals to the brain), or conductive (caused by a mechanical problem involving the eardrum and the three tiny bones of the middle ears), or a combination of both.
- Otitis media - commonly called glue ear, which is associated with cleft palates

2.4. JOINTS

- Bone and joint problems can include abnormality to ends of long bones, double jointedness (which can result in dislocated joints), joint pain and early onset of osteoarthritis.
- Scoliosis - curvature of the spine.
- It is most important to note that this is a progressive chronic condition, and the child will experience days when their joints are far worse than other days.

3. CARE OF A CHILD WITH STICKLER SYNDROME

3.1. GENERAL

- It is important to note that the symptoms and severity of Stickler syndrome vary considerably from patient to patient, even within a family, and can be difficult to diagnose. Most of the children affected by Stickler syndrome may look perfectly normal and this does mean that some teachers may disregard problems, especially if the child desperately wants to 'appear normal' to his or her friends.
- Although intelligence is normal, patients of school age may face considerable educational difficulties because of visual and/or hearing impairment.
- Some children with Stickler syndrome may need to be educationally assessed so that they will not become academically disadvantaged. If the child has not been statemented it may be advisable to seek an assessment.
- It is important for the child to be allowed to integrate in the class and school, but care of the eyes and joints **must** be taken into consideration.
- Encourage the child to become independent and to be responsible for his or her own property, especially spectacles, hearing aids and other aids like magnifiers, etc. However this will depend much upon the age of the child.

3.2. CLASSROOM MANAGEMENT

- A teacher should try not to stand in front of a window when talking to the class. Children with a visual impairment have difficulty in seeing when looking directly into bright light. It also makes lip-reading extremely difficult for those children who rely on this method of communication.
- Make sure that the child is looking directly at the teacher when instructions are being given to the whole class.
- When giving any instructions to the child in a one-to-one situation the teacher should always sit or stand on the child's 'good' hearing or vision side. Some children will have no peripheral vision.
- Teachers should speak directly to the child when they are giving one-to-one instruction, and make certain that the child understands what is being said. Ask discreet questions to ensure that the child has fully understood. When asked, some children will nod their head indicating that they have understood what they are being told as they do not want to admit to not hearing or understanding.

- When giving out homework, it would make good practise not to leave it until the end of the lesson or when the bell has sounded and everyone is rushing around and packing their bags. Write the homework on the blackboard, and make sure the 'Stickler' child has understood what is expected and when the homework needs to be completed.
- Make certain that any outside visitors or supply teachers are aware of the child's needs, especially on visits to a swimming pool. If the hearing aids or contact lens are removed then that child will not be able to see or hear properly and will need extra support.
- Be especially vigilant to the child's needs and abilities during school trips, particularly those that include overnight stays.

3.3. ABSENCES FROM SCHOOL

- Most children with Stickler syndrome will have numerous hospital appointments and admissions. Although parents/carers try to make appointments during school holidays, this is not always possible for a child with Stickler syndrome who needs regular check-ups and treatments. These appointments can have a detrimental effect on the child's education. Therefore it is important that the correct level of extra support is offered, and this needs to be monitored so that the child catches up without too much stress.

3.4. FATIGUE

- Children with Stickler syndrome are prone to chronic fatigue and this should always be remembered within the class environment as well as when the child is on field study visits, etc
- Some children may appear clumsy for various reasons. Please see details within the following headings.

All the points in this booklet MUST be re-emphasised each academic year as the child changes class/school and every teacher involved with that child should read this booklet - more copies are available free of charge from the SSSG. When supply teachers are used to cover absences amongst permanent members of staff, the Stickler child's needs should be highlighted.

3.5. VISION

- Teachers should always be aware that children with Stickler syndrome might not see clearly. For example there may be parts of their vision missing, and the child may appear to be clumsy because of poor depth perception. Others may have lost some of their field of vision, causing the child to stumble over obstacles such as waste paper bins, school bags left on the floor, or to walk into half opened doors. There may also be a presenile cataract present, which will make vision hazy and some colours like, blue, black, maroon and brown undistinguishable.
- Fragmented vision can cause problems, particularly if the child is unaware of a problem. Some tasks will take longer as the child tries to locate the next row of figures, next line, picture, etc so the child may be assessed as 'slow'.
- As vision fluctuates considerably according to lighting, there can be confusion about the extent of visual impairment. Good lighting in corridors and cloakrooms could prevent unnecessary accidents. Within budget constraints try to provide good lighting that avoids glare.
- Many children have reading as well as distance spectacles, and some may need to wear sunglasses when out in bright sunshine. Some may also need to use magnifiers. If the child has been prescribed spectacles, please ensure that they are worn, as children often don't wear them, as they do not wish to appear different.

- Even if the child has spectacles to help vision, the child should be near the front of the class so the teacher can see if he or she is experiencing any difficulties.
- Other children may wear contact lens or lenses, and the question concerning the removal of lens for any activities should be discussed with the parent. For Health and Safety reasons, and to avoid possible legal repercussions, teachers should not be encouraged to re-insert the lens, but the child concerned should be encouraged to carry a lens case for the lens to be housed for safe keeping.
- Select the appropriate typeface for all written material and, if necessary, have worksheets photocopied on to yellow paper - a colour that is easier for people with a visual impairment to read from.
- If a child complains of any loss of vision, however small or of seeing floaters, flashing lights, 'spiders' or 'black rain' descending before their eyes they **MUST** be taken very seriously. These could be the signs of an early retinal detachment. The parent/carer should be informed **immediately** and encouraged to take the child to the nearest hospital with an ophthalmology department, preferably one with a vitreo-retinal surgeon. If the family are members of the SSSG they would have been issued with a 'sight-card' to present at the hospital. It is vital that the help of an ophthalmic surgeon is sought. **Failure to do so could result in permanent loss of vision. Make sure that every member of staff is aware of the importance of eye disturbance.**

3.6. HEARING

- Children with a hearing loss find it difficult to take notes AND listen to what is being said. Therefore some facility should be made available for the child who experiences a problem, especially for children in secondary school or above.
- In a recent survey carried out by the Stickler Syndrome Support Group it was found that 11% of children with Stickler syndrome also suffered from tinnitus. This can affect their comprehension of lessons, and cause them to appear clumsy because of an inner ear balance problem.
- If the child wears behind-the-ear or in-the-ear hearing aids, please make sure that they are switched on and to the correct setting. This is especially important for younger children who tend to 'play' with the aid.
- Even if the child has hearing aids to help with hearing, the child should be near the front of the class so the teacher can see if he or she is experiencing any difficulties.
- Do not place the child too near to background noise, an open door or window, a piano, etc. Problems may be experienced for example at assembly and during music lessons. Children with a hearing impairment also experience difficulties during class discussions and find it difficult to follow them. Always make sure that the child is able to participate.
- If the child wears a radio aid be sensitive to the problems of transferring the microphone to different teachers, and the teacher taking assembly may need to wear the microphone. Problems transferring the microphone to different teachers may be more evident at secondary school level. Children are very conscious of being 'different' and if this is not handled with sensitivity it can become an issue for some children.

3.7. JOINTS

- Most children with Stickler syndrome suffer from joint pain and any 'Stickler' child complaining of pain should be taken seriously. Some children experience low-grade pain constantly, and this should be discussed with the parent/carer and not taken as an excuse to 'opt' out of any activity. Others will have good days and bad days.
- Children with Stickler syndrome are prone to hypermobile joints, which tend to dislocate easily. The most common joints that dislocate are the elbows, knees and shoulders. It is necessary to liaise with the parent/carer to determine the level of PE that is appropriate so that the PE curriculum can be modified to suit the Stickler child.

- Some children with Stickler syndrome experience stiff painful joints and may find it difficult to hold a pen and may use an easy grip penholder. Others may find it difficult to take notes and any difficulties should be discussed with the parent/carer.

3.8. PSYCHOLOGICAL

- The teacher will need to discuss, confidentially with the parent/carer, how much the child knows about his or her medical condition and how they feel about it. Any concerns that the child may have or discusses with the teachers should in turn be discussed confidentially with the parent/carer and an agreement reached on how best to deal with them.
- It may be possible during a 'Show and Tell,' 'Circle Time,' or a PSHE (Personal, Social and Health Education) session for the child to talk about what it is like to have Stickler syndrome or about one aspect of the condition e.g. sight, hearing, joints, coping with hospital visits, etc. It would also be a good idea to discuss with the parent/carer and ask how they would like the subject approached. The Stickler Syndrome Support Group will always send a batch of booklets for other members of the class, if this is felt appropriate. A school visit may also be arranged, if appropriate.
- Some children with Stickler syndrome, especially those who have a vision and hearing loss, mobility problem or speech impairment may be subject to bullying. The teacher should be aware of a change in the child, and the child should be encouraged to discuss issues with the form teacher as they arise.
- Some children who cannot hear or see properly or fall behind academically can become disruptive in class. Behavioural problems are not part of the condition, but can be caused by the difficulties the child experiences both at school and at home, particularly when there is a parent and siblings with the condition.
- In some families the condition may have gone undiagnosed for two to three generations, leaving in its wake a trail of emotional baggage, and this too must be considered when assessing the psychological needs of the child. It is not unusual when one member of a family is diagnosed for between 3 and 8 other members of the family, over three generations, to be diagnosed, on the same day.

3.9. PHYSICAL EDUCATION AND SWIMMING

- Sports and swimming can present a dilemma for the child as well as the school, and the level of activity should be discussed fully with the parent/carer. For example a bang to the head can trigger a retinal detachment, joint pain or hypermobile joints can make walking difficult. Although swimming is generally a good exercise, participation in this sport should be discussed with the parent/carer, especially diving, which is not recommended if the retina is weak. Make sure that PE teachers and other outside instructors are aware if the child wears contact lenses or hearing aids.
- Children with Stickler syndrome should generally be discouraged from playing contact sports. However this is an issue that should be discussed with the parent/carer and the ophthalmologist or rheumatologist caring for that child. If they are allowed to do any contact sport make sure the child is not wearing contact lenses or hearing aids, and if they are removed be aware that the child may be severely visually or hearing impaired without them.
- As a general rule the emphasis should be on gentle exercise, although it should always be remembered that joints that are overused will be painful the following day.

3.10. MEDICATION

- Children with Stickler syndrome need frequent medication, and although it is appreciated that most schools have adopted a 'we do not administer drugs' policy, some drugs are vital

to preserve sight. After discussions with the parent/carer perhaps this policy could be modified in the case of a child who needs medication for eye disorders, ear infections and joint pain.

3.11. ESTABLISHING A HOME-SCHOOL PARTNERSHIP

- Teachers should establish an open dialogue with the parent/carer so that concerns or queries regarding the child's health or academic progress can be discussed as difficulties arise, rather than waiting until a consultation evening. It is appreciated that as the parent/carer may work this can be difficult, but a home/school link book may be an alternative option.
- The school should also be aware of any other members within the family who have the condition, as the wider aspects of the condition may affect the child's progress at school. This may be especially true if the main parent/carer also has the condition, is disabled (immobile or blind), or may need to undergo surgery.

3.12. FURTHER EDUCATION

As children with Stickler syndrome go on to Further Education other special needs may arise. Further Education warrants a booklet of its own, and is therefore beyond the scope of this general booklet.

4. ABOUT THE SSSG

The Stickler Syndrome Support Group is the organisation that provides information and support for people affected by Stickler syndrome, carers and professionals. A series of booklets has been produced by the group on various aspects of the condition. These are:

- Stickler Syndrome What Is It?
- About the SSSG
- Stickler Syndrome - a Diagnostic Aid of Professionals
- Eye Involvement Within Stickler Syndrome
- Joint Involvement Within Stickler Syndrome
- Understanding genetics Within Stickler Syndrome
- Genetic Testing for Stickler Syndrome
- Cleft Involvement Within Stickler Syndrome
- Speech Therapy Within Stickler Syndrome
- Hearing Loss Within Stickler Syndrome

If you would like a copy of any of these booklets, further copies of this booklet, or have any further queries, please contact the SSSG, details overleaf.

Note: This booklet is not intended to be the definitive guide for teachers and youth workers working with Stickler syndrome children. If teachers and parents would like to contribute or offer any advice, please contact the SSSG so that we can take your comments on board. Thank you.

5. HOW YOU CAN REACH US

For further information write to the address below or contact:

**WENDY HUGHES
FOUNDER AND HON PRESIDENT
TEL/FAX: 01903 785771
E-MAIL: info@stickler.org.uk**

OR

Visit our website at www.stickler.org.uk

**Registered Office:
Stickler Syndrome Support Group
PO Box 3351
LITTLEHAMPTON
BN16 9GB**



Text © Wendy Hughes and the Stickler Syndrome Support Group.
All rights reserved. No part of this publication may be reproduced
or transmitted, in any form or by any means, without permission.