# **PUPIL INFORMATION**

**SURNAME:**  Click or tap here to enter text. **FIRST NAME** Click or tap here to enter text.

**DATE OF BIRTH:** Click or tap to enter a date.

**SEX:** Male[ ] Female[ ] Other[ ] Click or tap here to enter text.

**GENDER:** Male[ ] Female[ ] Other[ ] Click or tap here to enter text.

**MEDICAL CONDITION(S):** Ectodermal Dysplasia, Click or tap here to enter text.

**ALLERGIES:**  Click or tap here to enter text.

**MEDICATIONS (REGULAR):** Click or tap here to enter text.

# **CONTACT INFORMATION**

**Mother’s Name:** Click or tap here to enter text.

**Mobile #:** Click or tap here to enter text.

**Home Phone #:** Click or tap here to enter text.

**Work Phone #:** Click or tap here to enter text.

**Email:**Click or tap here to enter text.

**Address:**Click or tap here to enter text.

**Father’s Name:**Click or tap here to enter text.

**Mobile #:** Click or tap here to enter text.

**Home Phone #:**Click or tap here to enter text.

**Work Phone #:** Click or tap here to enter text.

**Email:** Click or tap here to enter text.

**Address:**Click or tap here to enter text.

**AUTHORISED EMERGENCY/ALTERNATE CONTACT**
**Name:** Click or tap here to enter text. **Relationship to child:** Choose an item.
**Mobile #:** Click or tap here to enter text.

# **MEDICAL CONDITION: Ectodermal Dysplasia. Specific type:** Ectrodactyly Ectodermal Cleft (EEC)

Ectodermal Dysplasia is a rare genetic condition that affects an individual’s temperature control, teeth, ears, eyes, skin, hair, nails, nasal passages, respiratory tract, muscles, behaviour, physical appearance, emotional wellbeing, and a host of other challenges.

The arrangements set out in this document are intended to assist the child, their parents, and the school in achieving the least possible disruption to school life and to maximise their education by making appropriate provision for their medical requirements.

A full explanation of how Ectodermal Dysplasia affects individuals is set out at the end of the document.

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# **DESCRIPTION: EEC SYNDROME**

EEC syndrome, one of the rarer 170 conditions has symptoms that vary greatly from one person to another. Affected individuals often have abnormalities affecting the limbs including ectrodactyly, a condition in which part or all of the central digits (fingers or toes) are missing. Ectrodactyly often affects the middle fingers or toes, but can present differently in different people (or be absent altogether). A groove or gap in the upper lip (cleft lip) and a groove or gap in the roof of the mouth (cleft palate) may also occur.

Most cases of EEC syndrome are caused by mutations of the TP63 gene and are either new (spontaneous) mutations or are inherited as autosomal dominant disorders.

Skin, hair, teeth, and sweat glands are commonly affected. Affected individuals may have dry, discoloured (hypopigmented) skin. The skin may also be itchy. Individuals tend to be fair skinned and have sparse, coarse, slow-growing scalp hair. Eyelashes or eyebrows may be sparse or absent.

Additional symptoms can include slow-growing, thin, malformed nails and missing, malformed or underdeveloped teeth. Tooth decay is common and often severe. Tooth enamel may be abnormal. Some individuals experience reduced activity or absence of certain glands including the sweat, salivary, and small oil-producing glands. Abnormality of the sweat glands can lead to a reduced ability to sweat, which can be associated with heat intolerance and fever. Abnormality of the salivary glands can lead to dry mouth.

Some individuals have eye issues including abnormalities of the tear (lacrimal) ducts that can cause frequent tearing, increased susceptibility to eye infections and chronic inflammation of the delicate membranes that line the inside of the eyes, potentially causing vision impairment. Additional abnormalities affecting the eyes can occur including sensitivity to light (photophobia), corneal ulcerations, inflammation of the cornea, and inflammation of the eyelashes and eyelids.

**Click or tap here to enter text. EEC SYNDROME PRESENTATION**
*For each section, describe the impact on the anatomy and how it presents in your child. Save how it should be managed for the next section, treatment plan. Add/remove anatomical areas as appropriate for your child.*

* Choose an item. Click or tap here to enter text.
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# Choose an item.Click or tap here to enter text.

# **TREATMENT PLAN**

All staff should be aware of Click or tap here to enter text.emergency plan.

Specialist subject teachers (especially sports teachers), supply/cover teachers, recess and lunch yard duty staff must be fully informed. This Healthcare Management Plan must be brought to the attention of all new members of staff, supply teachers and assistants.

 **OVERHEATING SIGNS**

* Reddening ears
* Reddening face
* Lack of concentration, lethargy, dreamy, uncooperative, fidgeting, disruptive, clumsy, badly behaved.
* Nausea
* Headache
* Seizure (rare).

# **PREVENTIVE ACTIONS TO KEEP Click or tap here to enter text. COOL:**

* Always ensure that Click or tap here to enter text.  is not seated next to windows or in direct sunlight in the summer months.
* A drink of water:Choose an item. *must be able to access cool water whenever*Choose an item. *needs it*. This is one of the easiest waysChoose an item. can regulate Choose an item. temperature.

Click or tap here to enter text. drinks a lot of water each day (which means Choose an item. might need to go to the toilet more often than other children).

* Ensure Click or tap here to enter text. wears Choose an item. hat and cool vest if outdoors between 10am and 3pm.
* Ensure Click or tap here to enter text. has permission to access an air-conditioned environment at any time of the day, including breaks.

**OVERHEATING TREATMENT**

* Remove any outer clothing.
* Ensure Click or tap here to enter text.  is taken to an air-conditioned room.
* Encourage Click or tap here to enter text. to put Choose an item. cool vest on over Choose an item. uniform. The vest should be rinsed in cold water and rung out…it may be used immediately or kept in a fridge.
* Encourage Click or tap here to enter text.  to sit down in a cool place.
* Encourage Click or tap here to enter text. to remove shoes/socks.
* Put feet on a cold gel pack.
* Offer Click or tap here to enter text. Choose an item. spray bottle to mist Choose an item. face with water.
* Encourage Click or tap here to enter text.  to drink cold water.
* Place cooling necktie around Choose an item.  neck.
* Wet Choose an item.  hair, neck and uniform if Choose an item. is not cooling down or Choose an item. mood/ behaviour is deteriorating.

If Click or tap here to enter text. mood/cooperation have deteriorated, Choose an item. likely needs to cool faster than Choose an item. has been to date. Please wet Choose an item. clothes or suggest a cold shower if Choose an item. will allow you. If Choose an item. is refusing and you are concerned about Choose an item. temperature, please contact Choose an item. immediately. **If you are unable to contact us or are concerned for her safety, please call an ambulance**.

If Click or tap here to enter text. is not co-operating or not cooling down, please call us, and bear in mind if Choose an item. is overheating it can take 1-2 hours to cool down to a satisfactory level.

**FURTHER SIGNS OF HEAT EXHAUSTION/HEATSTROKE ARE:**

* Headache.
* Dizziness.
* Confusion.
* Loss of appetite.
* Nausea.
* Pale skin.
* Cramps in arms,
legs or stomach,
* Rapid pulse and breathing.
* High body temperature.
* Deterioration in level of response.

Severe heatstroke if left untreated can be fatal.

If you are concerned about Choose an item. temperature, please contact Choose an item. immediately. **If you are unable to contact us or are concerned for her safety, please call an ambulance**.

|  |  |
| --- | --- |
| **FEBRILE SEIZURE:** **\****This has not happened to* Click or tap here to enter text. *before but could.*   | **ACTIONS:** **Do not leave** Choose an item. **alone and do not put ANYTHING in** Choose an item. **mouth.**   |
| If Choose an item. becomes too hot,Choose an item.  may have a seizure and become: * Unconscious.
* Unresponsive.
* Fall / Collapse.
* Stiff followed by rhythmical jerkingof limbs.
* Seizure ceases.

  | * Call 000.
* Note time.
* Loosen clothing.
* Put something soft under Choose an item. head.
* Do not restrict Choose an item.  movements.
* Move any surrounding objects.
* Monitor Choose an item.  breathing.
* Call parents.
* Allow Choose an item. to rest in the recovery position.
 |

# **ROUTINE SCHOOL TREATMENT**

# **SPORT/PE**

Click or tap here to enter text. must not be prevented from taking part in sport and should be actively encouraged to do so, but care must be taken regarding Choose an item. temperature. If Choose an item. is hot or cold Click or tap here to enter text. must be permitted to stop.

Click or tap here to enter text. must have a water/juice bottle with them at all times, and if doing field sports, it may help to have a bucket of water close by.

# **BREAK/LUNCH TIME**

Click or tap here to enter text. should be encouraged to go outside each day for fresh air. However, on hot/sunny days Click or tap here to enter text. must be encouraged to play in a shaded area and permitted to go back inside if Choose an item. is struggling with Choose an item. temperature, equally in the winter months’ encouragement to go out but permission to return if Choose an item. become too cold. It is not unusual for a child affected by ED to play outside in the winter without a coat; this is because they have become too warm in the classroom. However, they must take their coat outside ready to put on when they begin to get cold.

# **EYE IRRITATION**

When Click or tap here to enter text. eyes become sore, there are a number of possible causes including dryness, irritation related to pollen, some foreign object in Choose an item. eye(s) or infection.

Click or tap here to enter text. is very photophobic, so it is important that Choose an item. Has access to Choose an item. wraparound polarised sunglasses whenever Choose an item. requires them. Choose an item. must also combine these with a hat when outdoors for extra protection from sunlight.

# **FIRE DRILLS/EVACUATION**

Please assign someone to pay particular attention to Click or tap here to enter text.should an evacuation outdoors occur. Any walk or wait in the heat for any period can lead to Click or tap here to enter text. overheating quickly, so Choose an item. must be carefully monitored.

# **EMOTIONAL WELL-BEING**

Due to Choose an item. appearance and cooling requirements children often notice Click or tap here to enter text. differences quickly. Like most children, Click or tap here to enter text. does not like Choose an item. differences being pointed out. We encourage Choose an item. to answer however Choose an item. feels comfortable, but sometimes we provide some guidance as Choose an item. is still learning what is helpful. We worked on sharing a response that there is nothing *wrong* with Choose an item., and that Choose an item. was born this way, and that we are *all* different and that’s ok.  **It is important to us that** Choose an item. **maintains confidence and is supported when difficult conversations occur, as much as possible**. We know that the school can help tremendously by being aware of Choose an item. needs.

For a visual aid to better understand the temperature control problems experienced by individuals with ectodermal dysplasia, please refer to the image on the left.

The illustration depicts both an adult without ectodermal dysplasia and a child with ectodermal dysplasia at rest temperatures.

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ECTODERMAL DYSPLASIA FACT SHEET

**What is Ectodermal Dysplasia?**

Ectodermal Dysplasias comprise a diverse group of genetic disorders characterised by defects in the development or function of various ectodermal tissues, including hair, nails, teeth, skin, and glands. These disorders can affect additional parts of the body, such as the eyes, throat, skin, ears, and more. The term "ectoderm" refers to the earliest cells in a developing embryo that eventually give rise to teeth, hair, nails, and sweat glands, among other structures.

The term "dysplasia" indicates a deviation from the typical pattern of growth. Therefore, Ectodermal Dysplasia is a descriptive term denoting structural changes in body parts developed from the ectoderm.

With over 180 different types of Ectodermal Dysplasias identified, each syndrome presents a unique combination of symptoms, ranging from mild to severe. The key features of these disorders must exhibit abnormal functioning for a condition to be classified as an Ectodermal Dysplasia. The specific abnormalities and affected areas vary among the different types of syndromes. Ectodermal Dysplasia is a result of a genetic mutation passed from parent to child. In some cases, the genetic mutation occurs spontaneously in the affected person.

**Ectodermal Dysplasia Symptoms**

Each type of Ectodermal Dysplasia usually involves a different combination of symptoms, which can range from mild to severe, such as:

* Absence or abnormality of hair growth.
* Absence or malformation of some or all teeth.
* Impairment in the development of many glands, especially sweat glands, but also salivary glands (make saliva),
* lacrimal glands (make tears), mucous glands and the breasts.
* Lack of the ability to sweat causing overheating.
* Too little production of tears and other protective secretions of the eyes. This can make them sensitive and even painful.
* Reduced production of mucus in the airways, that leads to chest infections and – in those exposed too often to smoke or dust – to chronic lung damage (emphysema).
* Impairment or loss of hearing.
* Nasal blockage due to a build-up of secretions.
* Frequent infections due to immune system deficiencies and, in some cases, the inability to keep bacteria from entering the body through cracked or eroded skin.
* Less effective barrier properties of the skin, airways and gut leading to infections and to allergies (e.g. asthma, eczema and hay-fever).
* Respiratory problems: not only asthma and chest infections but also, in those who smoke or are exposed to dust, a severe form of chronic chest disease (may be diagnosed as emphysema or as pneumoconiosis).
* Absence or malformation of some fingers or toes.
* Cleft lip and/or palate.
* Irregular skin pigmentation.

In addition to the above individuals affected by Ectodermal Dysplasia may have:

* Sensitivity to light.
* A lack of breast development.
* Psychological challenges due to changes in physical appearance.

Individuals affected by Ectodermal Dysplasia may face a lifetime of special needs. These can include:

* Dentures at a young age with frequent adjustments and replacements.
* Osseo-integrated dental implants.
* Special diets to meet dental/nutritional needs.
* Air-conditioned environments.
* Wigs to conceal the lack of hair and scalp conditions.
* Creams or devices to protect from direct sunlight.
* Respiratory therapies for asthma and infections.

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