Hypohidrotic Ectodermal Dysplasias (HED) and Heat Balance

Heather Lunt,

Extreme Environments Laboratory, University of Portsmouth.

Human deep body temperature is maintained at around 37ºC. It is not a constant temperature; it can fluctuate by as much as a degree, being coolest in early morning and warmest during late afternoon and early evening. Maintenance of deep body temperature is a dynamic balance between heat production and heat loss. Heat production from physical activity or exposure to hot environments and heat loss when exposed to cold conditions is superimposed on this daily variation. As with most research, these facts are based on healthy adults (mostly male populations) as little research has been completed with other groups, including Hypohidrotic Ectodermal Dysplasias (HEDs), so the information provided draws upon the basic principles of heat exchange and the information we have regarding the body temperature regulation of HEDs.

The mechanisms of heat exchange enable heat balance to be maintained, preventing the deep body temperature from rising too high (hyperthermia, deep body temperature in excess of 39-40ºC) or cooling too much (hypothermia, a deep body temperature of 35ºC or less). In HEDs some of these mechanisms are compromised, primarily preventing loss of excess heat, leading to elevated deep body temperatures. This article will highlight the avenues of heat loss and describe strategies for heat loss in HEDs.

Heat production, primarily through the chemical process that occurs within our bodies to maintain life (metabolism), provides energy for the body to grow, repair and perform work (exercise or activity). The remainder is released as heat, which can increase deep body temperature. Heat exchange between the environment and humans occurs via the physical routes of convection, conduction, evaporation and radiation (Figure 1).

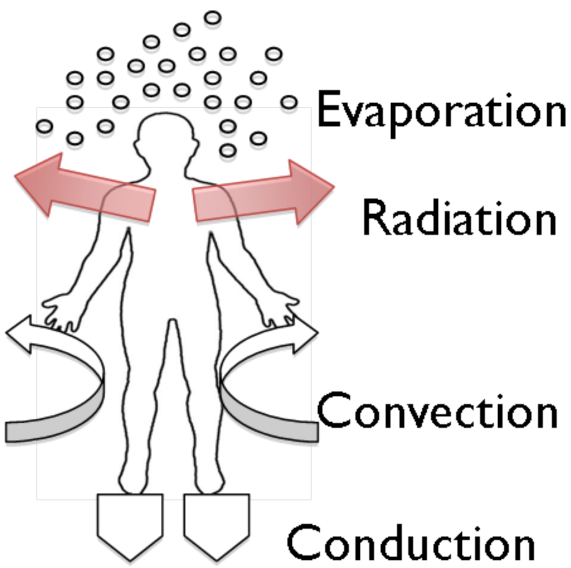

Figure 1. Avenues of heat loss

To take advantage of these means of cooling, different strategies can be used. Cooling using an ice vest, cold water immersion and contact of the feet with a cold floor are examples of conduction, which is the direct transfer of heat by contact of the body with a means of cooling. In contrast, convective heat loss occurs by rapid movement of air continuously replacing warmer air surrounding a body; standing close to a fan is a good example of convective heat loss. Radiation occurs by emitting heat energy into the environment; we can detect this using infrared imaging (Figure 2). Evaporation is the primary mechanism for heat loss particularly during exercise and/or in hot conditions. Evaporation of water from the skin or airway cools the body as the heat is transferred to the environment. For HEDs, evaporative heat loss from sweating is limited, placing greater reliance on radiation, conduction and convection heat loss. However, sweat can be substituted by spraying the body with water then allowing the spray to evaporate from the hot skin surface, further cooling can be enhanced by increasing convective heat loss by using fans.

Heat can be transferred to and from the body by these routes; the relative contribution to heat loss from each of these routes depends on the amount of activity undertaken and the environment the person is in. Increased air temperature, for example during a warm British summer with an air temperature of approximately $25^{o}C$ – $30^{o}C$, reduces the effectiveness of heat loss by conduction, convection and radiation. Furthermore, when the air temperature exceeds body temperature these routes of heat exchange (conduction, convection and radiation) contribute to heat gain. When this occurs or sustained exercise is undertaken, evaporation of sweat and water from the respiratory tract is the only avenue to remove excess heat. Consequently, overheating may occur due to a combination of events, and may result when fever or physical activity is combined with the body naturally being at its warmest, in a hot environment with impaired sweat function. This may lead to heat illness, sometimes referred to as heat exhaustion or heat stroke. The signs and symptoms of heat illness include: high deep body temperatures in excess of 38 or 39 ^{O}C, vomiting, diarrhoea, impairment of mental function, fainting or more prolonged unconsciousness, convulsions and impaired sweating even in individuals with functioning sweat glands. In HED, such signs and symptoms can occur quickly during hot spells, during exercise or fever and action should be taken to prevent heat illness. Therefore, what are the best methods of staying cool or cooling in order to enable you or your child to be physically active?

Figure 2. A thermal image

- Take exercise or be active when the body is at its coolest.
- Take regular breaks during exercise.
- Adjust (cool) the environmental temperature, through the use of air conditioning and/or shade on sunny days.
- Maximize heat loss from evaporation, conduction, convection and radiation, this could be through the use of air conditioning, cooling garments, immersion of the feet or hands in cool water, a cool shower, whole body spraying and fanning. To minimally disturb activities, these strategies could be applied during breaks and/or at the end of the activity.
- Adjust clothing layers to allow convective heat loss. Clothing keeps us warm by trapping air close to the skin and reducing the air movement between the skin and the environment. To cool we take off layers of clothing or wear loose clothing.
- Drinking cool fluids regularly will help to keep the airway moist; it is likely to have a very limited reduction in deep body temperature, but can make you feel cooler.

This practical support is at present applied from studies of healthy populations and needs to be investigated with HEDs. It is only through trial and error and development of coping strategies that you have learnt what works best for you. Some of this will be based on the principles discussed here, but there may be strategies that work for you that have not been mentioned here. If this is the case, please get in touch with us so that we can build studies that test out these strategies and may yield practical and useful information for all.

Thus far, you can see that most of what has been said is general advice, not very specific to HED. We are looking to undertake research into HED to ensure that advice about cooling and physical activity is tailored to you, rather than using the generic advice given above. For instance, at the start of the article we discussed the variation in deep body temperature over the course of the day. There has been no investigation of the daily variation in deep body temperature in HED. Therefore, the first area of research needed is to establish the daily pattern of deep body temperature of HEDs; to enable us to answer the question when the body is at its coolest and warmest. Consequently, we will then be able to recommend when to be active and when activity should be avoided.

There is a small amount of research already completed, which studied post exercise cooling using ice vests, this was shown to be effective in reducing deep body temperature in HEDs following exercise in the heat, by cooling the body at a faster rate than without the vest. Using ice vests to increase conductive heat loss is one method of cooling we could use. There are other methods, which take advantage of the conductive, convective and evaporative routes of heat exchange (mentioned earlier), that we would like to investigate to help establish the best methods of cooling HEDs. Undertaking this research will enable us to recommend the most appropriate techniques for a variety of activities and temperature conditions.

We can only undertake these studies with your help. Firstly, we would like you to tell us about your cooling strategies and what works best for you or your child, and secondly, we are looking for volunteers to participate in our studies. Over the summer term we are seeking young volunteers from school age up to age of 29 to participate in a field based study to discretely monitor your deep body temperature throughout the day and night. Following that we hope to run two studies to
determine the most effective cooling strategies after a short bout of exercise in a warm environment.